

# Mania as An Initial Feature of Systemic Lupus Erythematosus : A Case Report

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## Abstract

The initial feature of psychiatric symptoms in patients with systemic lupus erythematosus (SLE) is rare and difficult in diagnosis. This 42 y/o female is a victim of bipolar I disorder, she has no family history of psychosis or autoimmune disease. At the age of 29, she began to complain of psychiatric symptoms. Poor sleep, auditory hallucination, irritable mood, racing thought, hyper-talkative were the major symptoms. She came to our psychiatric clinic for help and was diagnosed as bipolar I disorder according to the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition criteria. During the 12 years of follow-up, flare up of symptoms was not frequent; she was admitted to acute psychiatric ward for 3 times due to manic episodes. Last Nov. she suffered from acute manic symptoms for 1 week. She came to our psychiatric clinic for help and was admitted. Malar rash was noted and series of examinations were arranged. EEG and contrasted Brain CT scan were negative. Because of the positivity of antinuclear antibody and anti-double strand DNA, the existence of psychiatric symptoms, photosensitivity, and malar rash, she was diagnosed as SLE, 12 years after manic present. Prednisolone 5mg bid was started first and adjusted to 7.5 mg qd after discharge with plaquenil 200 mg bid, improvement in the mental state and activity index of SLE of this patient have been observed. ( J Intern Med Taiwan 2007; 18: 283- 286 )

**Key Words** : Systemic lupus erythematosus (SLE), Mania

## Introduction

Systemic lupus erythematosus (SLE) is a multi-system disease of unknown etiology<sup>1</sup>. Psychosis is a rare but dramatic manifestation of SLE. Psychiatric symptoms are rarely reported as an initial feature of SLE and it is very difficult to diagnose such cases.

In this report, a case of SLE presenting with symptoms of Bipolar I Disorder was presented. Twelve years after psychiatric symptoms present, malar rash

was noted and series of evaluation reveal the diagnosis of SLE by positivity of antinuclear antibody (ANA), anti-double strand DNA (dsDNA), malar rash, photosensitivity and psychiatric symptoms.

## Case report

This 42 y/o female is a victim of bipolar I disorder. At the age of 29, she began to report of psychiatric symptoms. Major presentations include poor sleep, auditory hallucination, irritable mood, racing

thought, hyper-talkative. She came to our psychiatric clinic for help since then. She was diagnosed as bipolar I disorder according to the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV)<sup>2</sup>. During the 12 years of follow-up, her major medications include lithium carbonate 300 mg BID and artane 2 mg BID. She led an uneventful life most of the time and flare up of symptoms is not frequent. She was admitted to acute psychiatric ward for three times, Feb. 1993, Aug. 1996 and Feb. 2001, respectively due to manic episodes within this period. Last Nov. she suffered from mood related auditory hallucination (some kids ask her to join the hide-and-seek game, some one want to rape her), decrease need of sleep, wasteful expenditures of money, cognitive dysfunction, elated mood, agitation, poor attention, flight of ideas and hyper-talkative for 1 week. She came to our psychiatric clinic for help and was admitted under the impression of bipolar I disorder, manic episode.

On admission, malar rash was found (Fig. 1). The extent of redness subsides after avoiding sun exposure by remaining indoors. Tracing the history, the symptoms of malar rash and photosensitivity were noted by herself and her friends for about 1-2 year.

Physical examination revealed a thin woman with slight anxious looking and stable vital signs. Her consciousness is clear with intact skin, no subcutaneous nodules, no lymphadenopathy and no abnormality of head, eye, ear, nose, and throat were noted. Chest examination revealed clear breathing sound and regular heart beat. Physical examinations of other systems are unremarkable. No substance abuse or alcohol dependence history. No family history of psychosis or seizure disorder, nor any autoimmune disease history.

The laboratory results were as below: WBC count  $7950/\text{mm}^3$ ; Hb 11.6 gm/dl; platelet count  $28.6 \times 10^4/\text{ul}$ ; ANA 1:320 (+) homogenous pattern; rheumatoid factor negative; anti-dsDNA 20X(+); anti Sm(-); anti SSA(+); anti SSB(-); C3 62.7 mg/dl (90-180 mg/dl); C4 26.6 mg/dl (10-40 mg/dl); anti-cardiolipin Ab IgM 47.37 MPL (<12.5), anti-cardiolipin Ab IgG negative;

anti-ribosomal p Ab 2.82 unit (<20 U, negative); HBsAg negative, anti-HCV Ab negative, GOT 93 GPT 112; daily protein loss below 50 mg; RPR negative; anti-HIV antibody negative; T3, T4, TSH within normal limits. Chest x ray negative, EEG and contrasted Brain CT scan findings were normal.

Because of the positivity of ANA and anti-dsDNA, the existence of psychiatric symptoms, photosensitivity, and malar rash, she was diagnosed as SLE according to the 1982 revised criteria for the classification of systemic lupus erythematosus<sup>3</sup>. Prednisolone 5 mg twice a day was started and adjusted to 7.5 mg per day with hydroxychloroquine 200 mg twice a day after discharge. After the steroid therapy, activity of SLE seemed to get recovery, C3 value rose to 74.2 and 86.1 mg/dl, IgM anti-cardiolipin Ab 21.44 and 14.4 MPL respectively during the 4<sup>th</sup> and 11<sup>th</sup> week follow-up visit after discharge. Improvement in the psychological state of the patient has been observed during this period, no elevated mood, no irritable or talkative behavior, with normal sexual interest, motor activity and appropriate dress. Only dysphoric mood related to the stress of being SLE patient was noted. The psychiatric medications taken after discharge include risperidone 1 mg, luvox 50 mg hs and serenal 10 mg tid. Lithium is not necessary due to the improvement of psychological sta-

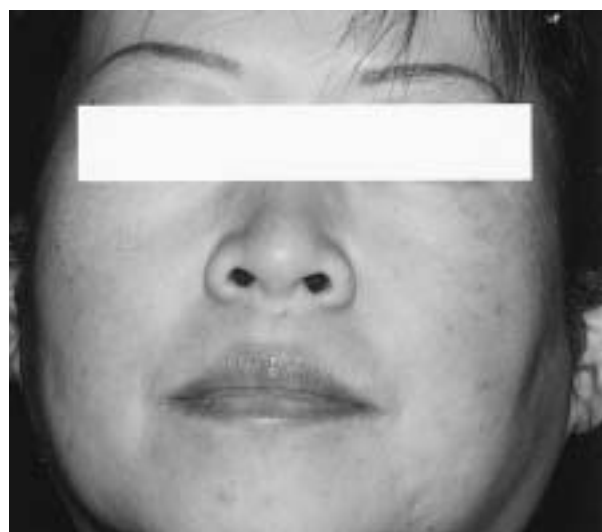


Fig.1. Malar rash of this patient

tus. The patient is under regular follow-up for around 5 months since SLE treatment and luvox was cancelled since last visit due to the continual improvements.

## Discussion

SLE is an autoimmune disease that involves many systems with variable manifestations. Psychiatric manifestations are rare but dramatic manifestation and are rarely reported as an initial feature of SLE. The involvement of Central Nervous System is an important manifestation of SLE. Neuro-psychiatric (NP) symptoms occur between 13% and 75% of SLE patients. Follow-up of 282 new-onset SLE Chinese patients after around 6.7 years, 65 patients (23%) had at least 1 NP manifestation and 50 (18%) developed NP damage. Cerebrovascular accident was the most common cause (35%), followed by seizure (20%), psychosis (12%)<sup>4</sup>.

The patients of SLE presenting with NP symptoms are very rare and diagnosing such cases is difficult. Feinglass et al. have reported neuropsychological manifestations as an initial feature of SLE in only 5 out of 140 patients<sup>5</sup>. Furthermore, in these patients, the initial events in SLE were rather neurologic than psychiatric. In this case, a 42-years-old woman presenting with symptoms of Bipolar I Disorder for about 12 year, malar rash was noted by the patient and her friend for about 1-2 year. This alarming symptom caught physician's attention on admission to psychiatric ward.

When psychiatric symptoms are present in SLE patients, they must be distinguished from other causes. Potential causes may not be excluded absolutely, and current or past illness and treatment may themselves cause neuropsychiatric manifestations. Secondary factors, such as drugs, metabolic abnormalities or infections, can also cause neuropsychiatric disturbances in lupus patients<sup>6</sup>. Psychiatric disorders may merely be reactive psychological disturbances due to the stress related to the impact of SLE itself and its treatment<sup>6,7</sup>. In this patient, no other CNS symptoms were found and EEG, contrasted brain CT scan showed negative results, no drug use, no

signs or symptoms of metabolic abnormalities or infection. Her diagnosis of psychosis is far ahead of the diagnosis of SLE; therefore, it will not be reactive psychosis related to SLE.

Since psychiatric disorder may be present in SLE patients, it raised a question of whether or not an auto-antibody screen is a useful investigation in psychiatric practice. An auto-antibody screen for SLE was performed on 296 patients admitted to acute psychiatric wards. Three cases (1% of those screened) of previously undiagnosed SLE were found<sup>8</sup>. Another study which tested ANA and antibodies to DNA in sera from 2121 psychiatric admission patients and from 500 sex- and age- matched controls, ANA were positive in 3% of patients and controls. Anti-DNA antibodies were found in 1% of both patients and controls<sup>9</sup>. Routine screen tests for the antinuclear and anti-DNA antibodies on psychiatric admissions thus would seem difficult in helping finding out more cases of SLE from these studies.

During the last 2 decades, the utility of antibodies to ribosomal P proteins (anti-P) in detecting NPSLE was examined. The association between psychosis, depression and anti-P antibody in SLE was supported by some but not all studies. An international meta-analysis study combined standardized data from 1,537 lupus patients contributed by 14 research teams showed the weighted sensitivity and specificity estimates for psychosis, mood disorder, or both were 27% (95% CI 14-47%) and 80% (95% CI 74-85%) respectively<sup>10</sup>. Therefore, anti-P antibody testing has limited diagnostic value for NPSLE. In this case, the negativity of anti-P antibody also showed that testing for anti-P antibody is not useful in identifying disease-mediated psychosis or mood disorder with enough certainty, since more than 60% of cases are negative.

The etiology and pathogenesis of psychiatric manifestations of SLE are mostly unknown. To which degree these psychiatric conditions of this patient can be explained as part of the disease spectrum of SLE or simply a coincidence, is a matter of debate. The

evidence of no mania symptoms and stable mood after treatment of SLE along with activity index of SLE recovered may support the diagnosis of lupus psychosis, although the follow-up period till now is less than 5 months only.

The diagnosis of affective disorder could be made readily according to the history and mental examination. SLE should also be considered in patients presenting with psychiatric symptoms when alarming symptoms developed.

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# 以躁症為初發表現的全身性紅斑狼瘡： 個案報告

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## 摘 要

全身性紅斑狼瘡是慢性的自體免疫疾病，以精神症狀為初發表現的全身性紅斑狼瘡個案相當少見。本文報告一位42歲女性，沒有精神疾病或自體免疫疾病家族史，在29歲時出現聽幻覺、失眠、情緒不穩、多話等症狀，而被診斷為雙極性情感性精神病。在十二年的追蹤觀察中，曾因為躁症發作住院三次。去年十一月因情緒高亢，活動量大，自大，意念飛躍等躁症症狀來求診而住進急性精神科病房。住院時發現有雙頰紅疹和光過敏的現象，經過一連串檢查後發現抗核抗體陽性、抗雙股DNA抗體陽性，腦電波和電腦斷層檢查並無異常。此病人在躁症出現12年後，因雙頰紅疹、光過敏、精神疾病、抗雙股DNA抗體陽性和抗核抗體陽性診斷為全身性紅斑狼瘡。經過治療後第三補體回升，精神症狀改善，病人繼續在門診追蹤中。