Increase Clinical Awareness of SLE Manifestations

翁孟玉

成功大學醫學院 助理教授

附設醫院 過敏免疫風濕科

Epidemiology

The rate of SLE varies considerably between countries, ethnicity, gender, and changes over time

- United States
  - The incidence : 7.6/100,000 in 1974
  - Prevalence : 53/100,000
  - Northern Europe : 40/100,000 people
  - Afro-Caribbean descent : 159/100,000
- Taiwan
  - The average annual incidence rate : 8.1/100,000
  - The prevalence : 67/100,000 in 2007
  - Woman : Men = 7:1

Child-bearing years ages women (20-40 y/o)
Poor prognosis of male patients with SLE

Clinical manifestations

SLE = The great imitators
SLE symptoms vary widely
Myriad symptoms, complaints
Inflammatory involvement that can affect virtually every organ

Constitutional Complaints

Fatigue : The most common complaint (80-100%)
Myalgia
Weight loss
Fever
Lymphadenopathy

Specific Organ Involvement

Raynaud's Phenomenon

Cold- or emotion-induced color changes of the digits of the hands and/or feet
16 – 40% of SLE patients
Sharply demarcated color change
3 Phases

Mucocutaneous

Oral ulcer : 12 – 45% of patients

The oral ulcers are usually painless
May be the first sign of lupus

Dermatological

Cutaneous manifestations of SLE include 3 American College of Rheumatology (ACR) lupus diagnostic criteria: malar rash, discoid lupus and photosensitivity

As many as 30% of sufferers have some dermatological symptoms
65% suffer dermatological symptoms at some point
30% - 50% : Malar rash (butterfly rash)
Discoid rash : alopecia
Photosensitivity
Cutaneous manifestations, not specific to SLE :

- Livedo reticularis
- Bullous lesions
- Vasculitic purpura

Neuropsychiatric Symptoms

Central and/or peripheral nervous systems
American College of Rheumatology (ACR) defines 19 neuropsychiatric syndromes in SLE
Cognitive dysfunction
Mood disorder
Depression
Mania
Psychosis
Cerebrovascular disease
Headaches
Seizures
Polyneuropathy

Cardiac

Serositis : pericarditis relatively common
Myocarditis : uncommon
Endocarditis : Libman-Sacks endocarditis involved either the mitral valve or tricuspid valve
Atherosclerosis
Pulmonary hypertension

Pulmonary

Serositis

- Pleuritis
- Pleural effusion

Lupus pneumonitis
Pulmonary hemorrhage
Interstitial lung disease
Pulmonary hypertension

Pulmonary emboli
Shrinking lung syndrome

Hematological

Leukopenia : WBC <4000/mm^3 X 2
Lymphopenia : <1500/mm X 2
Hemolytic anemia
Thrombocytopenia : Plat < 100,000
Antiphospholipid syndrome (APS)

Reproductive

Increased rate of fetal death *in utero*
Sontaneous aboriton
Pregnancy outcome appears to be worse in SLE patients whose disease flares up during pregnancy.

Musculoskeletal

Arthralgia : > 90% of patients at some time during the earliest manifestation
Arthritis :

- 65-70%
- Migratory and symmetrical
- Rarely erosive or deforming

Myalgia

Renal

Proteinuria
Lupus nephritis : glomerulonephritis
Renal involvement : 50% of SLE patients
Usually develops in the first few years of SLE
Renal biopsy

Gastrointestinal Tract

Mesenteric vasculitis
Bowel infarction
Pancreatitis
Colitis
Liver involvement
Protein losing gastroenteropathy

Diagnosis

Laboratory tests

ANA : mainstay of serologic testing for SLE
Screening tool

Subtypes of antinuclear antibodies :

- Anti-double stranded DNA ab (70%) : related to disease activity
- Anti-Smith /RNP ab

Antiphospholipid ab :

- LAC(lupus anticoagulant ab)
- Anti-cardiolipin IgM/IgG ab
- Anti-beta2 glycoprotien I ab
- VDRL

C3, C4
CBC/DC
U/A

ACR classification criteria
SLE : 4 out of 11 symptoms
Were established mainly for use in scientific research
It was not intended to be used to diagnose individuals and do not do well in that capacity
Clinical judgment

Clinical Course of SLE
The pattern that dominates during the first few years of illness tends to prevail subsequently
Disease activity come and go unpredictably

- The clinical course of SLE is variable
- May be characterized by periods of remissions and of chronic or acute relapses

結 語
SLE is a chronic systemic autoimmune disease
It can affect almost any organ system
Its presentation and course are highly variable
Disease activity characterized by periods of remissions and flares
In patients with suggestive clinical findings should raise further suspicion of SLE
Early diagnosis improved the outcome of SLE