Clinical Case: LUPUS
### Case Study

Annabel is a 20 year-old Asian-American female currently in her third year of college. She presented to the student health center complaining of four days of persistent fatigue and arthralgia in her hands and wrists. Annabel states she had a similar episode with fever during last term’s final exams but thought she was “run-down from studying.” Annabel denies any fever, weight loss, nausea, vomiting or rash but states she sometimes gets “red splotches” on her arms if she is in the sun too long. Annabel reports no acute medical history or regular medications.

#### Annabel at a Glance

<table>
<thead>
<tr>
<th>20 years old</th>
<th>Vitals upon exam:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Height: 5’2”</td>
<td>» Temp: 98.6°F</td>
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<tr>
<td>Weight: 108 lbs</td>
<td>» HR: 84 bpm</td>
</tr>
<tr>
<td>No acute medical history</td>
<td>» BP: 128/88</td>
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<tr>
<td></td>
<td>» Resp: 12</td>
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|  » Skin: No visible rashes or discoloration on exam |
|  » Joints: Some tenderness, but no appreciable swelling |

#### Is It Lupus?

Annabel’s presentation and history are nonspecific and may indicate one of several different diagnoses, including an autoimmune disease.

Systemic lupus erythematosus (commonly called SLE or lupus) is an autoimmune disease which produces widespread inflammation and tissue damage. Lupus may affect the skin, joints, kidneys, lungs, nervous system and other organs of the body. The presentation and course of lupus are variable, ranging from indolent to fulminant.

Lupus most often strikes people in their 20s and 30s, and is more common among Blacks and Asians. There is no known cause for lupus, but it is believed to be influenced by genetic, environmental and hormonal factors. Most patients experience flares, or periods where the disease is active, followed by periods where the disease is quiet, referred to as remission.

Several organ systems may be affected by inflammation caused by lupus:
- **Kidneys**: lupus can cause kidney damage and failure, indicated by itching, chest pain, nausea, vomiting and edema.
- **Brain**: lupus can cause headaches, dizziness, behavior changes, strokes or seizures. Patients may also experience memory or concentration problems.
- **Circulation**: lupus can lead to anemia, increased risk of bleeding or blood clots, as well as vasculitis. Lupus may also increase the risk for heart attack.
- **Bones**: lupus, as well as the steroids sometimes used to treat it, may increase the risk of avascular necrosis of bone. This is most common in patients’ hip joints.
- **Pregnancy**: women with lupus have an increased risk for miscarriage, preeclampsia and preterm birth.

#### SLE is the most common form of lupus, but other types exist:

1. **DISCOID LUPUS ERYTHEMATOSUS**
   - Causes a skin rash that does not go away

2. **SUBACUTE CUTANEOUS LUPUS ERYTHEMATOSUS**
   - Causes a skin rash on sun-exposed areas

3. **DRUG INDUCED LUPUS**
   - Can be caused by medications

4. **NEONATAL LUPUS**
   - A rare type of lupus that affects newborns

Lupus-related mortality is mainly caused by organ failure, such as the kidneys, infection or cardiovascular disease from accelerated atherosclerosis. Decreased mortality rates associated with lupus are a result of earlier diagnosis and improvements in treatment. But the Centers for Disease Control and Prevention estimates one-third of lupus related deaths occur in patients younger than 45 years.

While Annabel appears normal on exam and denies family history of autoimmune disease, her age, race, episodic arthralgia and photosensitivity raise your suspicion of lupus.
Making a Lupus Diagnosis

Sometimes called the “great imitator,” lupus can be difficult to diagnose because it mimics many other illnesses. According to CDC, underdiagnosis persists because presenting symptoms are often nonspecific.\(^1\) Initial presentation is often constitutional. The classic presentation of a triad of fever, joint pain and rash in a woman of childbearing age should prompt investigation into lupus.\(^2\)

Despite Annabel’s lack of fever upon exam, her report of fever during a prior episode of arthralgia and fatigue make you suspicious she could have experienced a prior flare.

The most important blood screening test for lupus measures antinuclear antibodies (called ANA). But a patient can have ANA and not have lupus. High levels of ANA are found in 98% of patients with lupus. However a number of other conditions also result in a patient having high levels of ANA. These conditions include other autoimmune diseases such as scleroderma, Sjogren’s syndrome and rheumatoid arthritis. ANAs can also exist in perfectly healthy women.\(^4\)

The ANA blood test is your initial, but vital, screening test. If your patient does have a positive ANA, further investigation is needed to make a diagnosis and referral to a rheumatologist is appropriate.\(^3\)

The American College of Rheumatology has outlined several symptoms which may present during initial onset. The presence of four of the criteria below is suggestive of the diagnosis:\(^3\)

- Rashes
  - Malar rash (butterfly rash)
  - Discoid rash
  - Rash on skin exposed to the sun
- Mouth sores
- Arthritis
- Lung or heart inflammation
- Kidney problems
- Neurologic problems
  - Seizures
  - Stroke
  - Problems with balance
  - Problems with vision
- Abnormal blood tests
  - Low blood cell counts
  - Positive antinuclear antibody
  - Other antibodies demonstrating an immune system problem

Look Out for the Lupus Triad
(in women of childbearing age):

1. FEVER
2. JOINT PAIN
3. RASH

Lupus Does That?

**Brain**
- Headaches, dizziness, behavior changes, strokes, seizures, memory or concentration problems

**Circulation**
- Anemia, increased risk of bleeding or blood clots, increased risk for heart attack, vasculitis

**Kidneys**
- Kidney damage and failure, indicated by itching, chest pain, nausea, vomiting and edema

**Pregnancy**
- Increased risk for miscarriage, preeclampsia and preterm birth

**Bones**
- Increased risk of avascular necrosis
Lupus By The Numbers

- Prevalence estimates range as high as 1.5 million Americans\(^1\)
- Lupus accounts for 14.5% of all rheumatic disease mortality\(^1\)
- Lupus affects 10 times as many women as men\(^3\)
- Mortality is five times higher among women with lupus than men with lupus\(^1\)

One-third of lupus-related deaths occur in patients younger than 45 years.

Referring to a Rheumatologist

Like many presentations of lupus, Annabel’s case is nonspecific. However, she is a young woman of Asian-descent, childbearing age with episodic arthralgia, fatigue and a photosensitive rash.

Annabel is clearly worried and asks how her sensitivity to the sun is relevant now, since she is not currently showing symptoms. You counsel her on how autoimmune diseases may affect different organ systems. You advise her to rest and take an OTC NSAID for pain relief as needed. You also suggest a consultation with a rheumatologist. Annabel asks if she can do anything until she is seen by the rheumatologist and you explain there are a number of things she can do herself before she meets with a rheumatologist:

- Keep careful track of any other symptoms she experiences
  - Paying special attention to anything which may act as a trigger of her symptoms
  - Keep a note of anything which makes her symptoms better or worse
- Use sunscreen and be mindful of sun exposure
- Return to the student health center with any additional symptoms or concerns before her appointment with her rheumatologist.

Lupus Referral Checklist

- Perform a complete physical exam
  - Carefully look for any unusual skin rash or discoloration
  - Carefully look for swollen joints
- Complete blood count
- Comprehensive metabolic panel
  - Urinalysis with microscopic exam
- Antinuclear antibodies results
- Document when ANA was measured
- Document timing of labs
- Document timing of flares
  - Knowing when labs were taken in relation to lupus flares may be important
- Timeline of any medications
  - What medications?
  - When did patient start and stop the medications?
  - When did patient experience flare ups in relation to medications?
  - Were any of the medications taken in combination? If so, when? Any flare ups?

As Annabel’s primary care provider, your role is essential to her long-term care and ensuring she stays informed and observant of any symptoms indicative of an autoimmune disease.

Citations


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