

YF Case Discussion Guide

Key Learning Objectives

- List the differential diagnosis for a systemic disease presentation
- Describe the key manifestations of disease on physical exam in a multisystem disease presentation
- Discuss the approach to the diagnosis of systemic lupus erythematosus
- Summarize questions for consultants in the inpatient setting
- Summarize socioeconomic issues and their impact on clinical outcomes
- List the guiding principles of cultural competency

Case Presentation

An 18-year-old Spanish-speaking Mexican female with complaints of nausea and vomiting as well as weight loss. Her parents, who speak a little English, provide most of the history.

The emesis is not related to food and consists of yellow fluid and food, no blood. There is no associated abdominal pain, diarrhea, constipation, hematochezia, or melena. She denies dysuria or hematuria. Her last menstrual period was normal and was approximately 10 days ago. Her review of systems is otherwise negative.

Approximately 2 weeks ago, she was given an oral antibiotic from an outside hospital for a presumed urinary tract infection. Shortly after the antibiotic was started, she developed symptoms of abdominal pain and fever. She continued to take the antibiotic for several more days, thinking it would get better with the medication. When symptoms progressed to significant nausea and vomiting, her family brought her to the hospital.

Her past medical history is significant for a healthy pregnancy delivered by cesarean section 2 years prior. There is no known family history.

What other questions should you ask about her personal and/or family history?

In a young female it is important to obtain additional personal and family history, including obstetric history (specifically miscarriage and/or premature births).

How can language barriers impact this patient's evaluation and care?

Please refer to the vignette "Sick and Tired of Being Sick and Tired." In the vignette, linguistic competence is the capacity of an organization and its personnel to communicate effectively, and convey information in a manner that is easily understood by diverse groups, such as this Spanish-speaking female and her family. Discuss groups in your community and the potential communication barriers they may face. Discuss how your organization, clinic, or hospital lower these barriers and how they could improve.

She denies use of alcohol, tobacco, or drugs and lives with her parents and the baby's father. None of them is documented as living in the United States. She did not graduate from high school and works cleaning homes.

She moved from Mexico with her parents when she was about 4 years old. Her parents and the baby's father are very supportive and they all provide childcare. Her father works in construction, her mother does not work outside of the home

and the baby's father has irregular jobs cleaning office buildings. Together, she and her baby's father make about \$1400 per month. She and her baby's father speak Spanish only; her parents and a cousin speak some English.

She does not take any prescription medications or over-the-counter medications regularly. One of her family members later brought the bottle of antibiotics she was taking for her presumed urinary tract infection. It was trimethoprim/sulfamethoxazole (Bactrim).

What kind of antibiotic is this? What conditions is this class of antibiotics associated with causing? How can the process of prescribing and dispensing antibiotics and other medications be improved to maximize the opportunity to achieve good outcomes?

Trimethoprim/sulfamethoxazole is a sulfonamide and has been associated with blood dyscrasias, Stevens-Johnson syndrome, and other drug allergies. It also has been associated with flares in patients with known SLE.

This is also an opportunity to discuss literacy issues that may have contributed to the progressive symptoms in this patient. Literacy in general is the ability to read and understand materials in a particular language. The National Assessment of Adult Literacy (NAAL) found that for Hispanics in America the average literacy scores for reading documents fell 14 points from 1992 to 2003. Individuals who spoke Spanish before starting school comprised only 8% of the sample for the NAAL, but comprised 35% of those who were rated below "basic" in prose literacy skills.

SOURCE: U.S. Department of Education, Institute of Education Sciences, National Center for Education Statistics. A First Look at the Literacy of America's Adults in the 21st Century.

Downloaded on 8/1/2011 from <http://nces.ed.gov/naal/pdf/2006470.pdf>.

Studies show that persons with low literacy skills are less likely to

- Seek and get preventive care
- Understand forms for informed consent
- Understand their children's diagnosis
- Understand medication instructions for themselves and their children and
- Be knowledgeable about the health effects of risks, behaviors, and diseases

(Berkman, N., DeWalt, D. et al., 2004)

Health literacy goes beyond reading and comprehending written material and is particularly pertinent in this case. The Patient Protection and Affordable Care Act defines health literacy as “the degree to which an individual has the capacity to obtain, communicate, process, and understand health information and services in order to make appropriate health decisions.” (Title V—Health Care Workforce, Subtitle A—Purpose and Definitions.) This patient may not have been able to read and understand medication information and warnings that are often given with prescriptions. In addition, health literacy impacts a range of facets of healthcare, including the ability to:

- Access information
- Recognize cues to action
- Access care
- Navigate institutions
- Complete forms
- Provide consent
- Communicate with professionals
- Provide information for assessment, diagnosis, & treatment
- Understand directions
- Follow regimens
- Advocate

SOURCE:Rudd, R.E. (2003). Empowering Disadvantaged Populations. [Electronic Slide Presentation]
Retrieved from http://www.hsph.harvard.edu/healthliteracy/overview_slides.pdf, on 7/22/05
Harvard School of Public Health, Health Literacy Studies

Issues of health literacy are further challenged by those patients who have limited English proficiency or are monolingual Spanish speakers, particularly given the lack of translated documents, bilingual staff, or interpreters who can assist with reading and reviewing documents with patients. Providing quality healthcare includes addressing the literacy and health literacy of patients as an aspect of linguistic competence.

Patients may prefer to get health information in different formats. As noted in the definition of linguistic competence, the goal is effective communication of important information that patients need to address their health. Thus, health information should be provided not only in the language patients prefer, but also at a literacy and health literacy level they can understand, using plain language (<http://www.nih.gov/clearcommunication/plainlanguage.htm>) and in a format that maximizes their learning.

Physical Exam

Vitals: Blood pressure **97/53**, heart rate **114**,
respiration rate 20, Sat 99% room air

Temp: **38.4 C (101.1 F)**

General: No acute distress

HEENT: Pale conjunctiva, no ulcers, no pharyngeal edema

Pulmonary: Clear to auscultation bilaterally

Cardiovascular: Normal heart sounds, no murmurs

Skin: No rashes

Musculoskeletal: No synovitis, full range of motion throughout

Where should one specifically look for ulcers within the oropharynx?

Aphthous ulcerations are generally painful and can be found on the buccal mucosa, tongue, inner lips, and sometimes on the hard palate. Oral ulcers due to systemic lupus erythematosus are generally not painful and can be found on the hard palate.

Data

Na **121**, K 4.7

Cl **98** HCO₃ **19**

BUN/Cr 22/1.0

Total protein **5.9**, albumin **2.0**

Calcium 7.0, CorrCa 8.6

T bili/ALT/AST/Alk phos 0.05/**63/147/29**

Lipase **128**

CPK 275

WBC **0.824**

48% Segs (ANC **396**)

12% Monos

40% Lymphs (absolute lymphocyte count **330**)

Hg/Hct **10.5/32.2**, MCV 84

Plt 72

PT/PTT/INR/fibrinogen **14.0/48.8/1.22/231**

Urinalysis

3+ protein

3+ blood

1+ ketones

No nitrates or LE

WBC 11–23

RBC 2–5

Hyaline casts 2–5

Granular casts 6–10

Cellular casts 2–5

Urine pregnancy negative

She is admitted to a general medicine team and placed in a room on a regular medical floor.

Since she is febrile and meets systemic inflammatory response syndrome (SIRS) criteria, the team treats her with antibiotics (vancomycin and piperacillin/tazobactam), while a source infection was investigated. Metoclopramide as needed for nausea was ordered.

The following day, more data are available for the team to review.

Blood cultures: No growth to date

Urine cultures: No growth to date

Hemoglobin 8.5, MCV 85

MRI abdomen with and without contrast

Small bilateral pleural effusions

No obstructing hepatobiliary, bowel, or pancreatic lesion is identified

Although her blood and urine cultures are negative she remains febrile with the same gastrointestinal complaints

What could be causing her fever at this point?

The students should be thinking about causes of fever other than infection, such as a drug reaction, malignancy, and connective tissue diseases.

What processes could be the cause of her active urinary sediment? How should this be evaluated further?

The students should be thinking about the initial steps in evaluating a urinalysis containing protein and blood. Proteinuria should be quantified with a spot urine protein to creatinine ratio and/or a 24-hour urine collection. Furthermore, the urine should be evaluated by microscopy to assess whether or not the RBCs are dysmorphic. Dysmorphic RBCs should prompt them to think about diseases originating from the glomerulus (as opposed to the genitourinary tract), such as lupus nephritis in this patient in particular.

The Hematology service is consulted to assist in evaluation of her pancytopenia.

They consider etiologies, including infection, malignancy, immune-mediated processes and nutritional deficiencies.

Hematology recommends ordering hemolysis labs (LDH, haptoglobin, peripheral smear, Coombs test), iron studies, vitamin B12 and folate levels. They also agree with the connective tissue disease and atypical infectious diseases workups already in place by the primary team. They do not recommend a bone marrow biopsy at this time.

The team initiates the workup above and also continues their workup for infectious etiologies, including parvovirus. The patient remains stable and the team decides to continue with antibiotics for the time being.

Additionally, the medicine team spins her urine and under microscopy, determines that the RBCs in her urine are dysmorphic.

Some of the additional labs ordered return:

C3 **22** C4 **<8**
ESR **65**

LDH **395**
Haptoglobin **<30**
Coombs +

Iron 114, 68%, transferrin 111

Folate 7.9 B12 928

HIV –
EBV PCR –
CMV PCR –

Spot urine protein to creatinine ratio estimates **0.7g of proteinuria/24 hrs**
24-hour urine collection in process

Within a couple of days of her admission, with the infectious workup negative thus far, the Rheumatology service is consulted. Their evaluation is done with a Spanish-speaking medical translator (unlike her initial evaluation).

Rheumatology Consult Service Physical Exam

Vital: Blood pressure 100/70, heart rate 98, respiration rate 14, sat 99% room air

Temp: 38.3 C (100.9 F)

General: Young female, lying in bed, in no acute distress

HEENT: pale conjunctiva, normal sclera, angular cheilitis, peeling lips, 2 large deep ulcers on hard palate anterior behind front teeth

Cardiovascular: normal heart sounds, no murmurs

Pulmonary: Clear to auscultation bilaterally

Abdomen: Soft, + bowel sounds, nontender, nondistended, no organomegaly, no rebound tenderness

Skin: Fixed livedo nonblanching lesions on palms (thenar) and finger tips

PICTURES OF ORAL ULCERS
FROM ACR IMAGE BANK TO BE
INSERTED

PICTURES FROM ACR IMAGE
BANK TO BE INSERTED

No digital ulcerations, no nail fold capillary changes

Musculoskeletal: No synovitis, full range of motion throughout

Neurologic: Cranial nerves grossly intact, reflexes and strength within normal limits, some delay in following simple commands as directed by the translator

How does the Rheumatology physical exam affect the case?

The Rheumatology consultation provided insight into the physical exam that was not appreciated previously. The patient was appropriately interviewed by a Spanish-speaking translator, which revealed her slightly delayed mental status. Additionally, her painless oral ulcers and vasculitic rash were appreciated.

Her ANA screening test by ELISA is positive, however the immunofluorescence pattern and titer are pending.

With the above clinical picture, including a completely negative infectious workup, new-onset systemic lupus erythematosus is the most likely unifying diagnosis.

What are the American College of Rheumatology Classification Criteria for SLE?

Please refer to the Lupus Initiative lecture series. Review with the students slide 8 of Dr. Mackay's lecture. Here, the 11 American College of Rheumatology (ACR) criteria are described and it is important to note that these must be attributed to SLE to be counted towards the classification. The criteria do not include some of the more commonly found manifestations, including fatigue and arthralgia, which are nonspecific by themselves; attribution to SLE can be challenging in the absence of other features.

How do you make a diagnosis of SLE and do patients have to meet 4/11 ACR criteria to have a diagnosis of lupus?

This is a good time to discuss with the students that systemic lupus erythematosus is a clinical diagnosis. Serologic tests alone do not make a diagnosis. The patient must have both the clinical history and physical findings, using serologic tests as supporting data.

Due to the relatively low sensitivity of the ACR criteria, many physicians do not rely solely on the criteria when making a clinical diagnosis of lupus. The example Dr. Mackay gives in her lecture is of a young, Hispanic female with months of fatigue, lymphadenopathy, + ANA consistent with SLE but not meeting ACR criteria.

What would you do if a translator was not quickly available?

One could point out that the essential clues in this case were in the laboratory studies. However, beyond the diagnosis, certain aspects of her clinical presentation are important to define. This patient's

level of understanding could be due to SLE, a language barrier, or other causes. This again underscores the importance of linguistic competence in health, which is defined as: The capacity of an organization and its personnel to communicate effectively, and convey information in a manner that is easily understood by diverse groups, including persons of limited English proficiency, those who have low literacy skills or are not literate, individuals with disabilities, and those who are deaf or hard of hearing.

Linguistic competency requires organizational and provider capacity to respond effectively to the health and mental health literacy needs of populations served. The organization must have policy, structures, practices, procedures, and dedicated resources to support this capacity.

Goode & Jones (modified 2009). National Center for Cultural Competence, Georgetown University Center for Child & Human Development. For a full definition, click here: <http://nccc.georgetown.edu/documents/Definition%20of%20Linguistic%20Competence.pdf>

This also demonstrates the need for another aspect of linguistic competence in care settings—the need for language assistance services. 43.1% of individuals who speak a language other than English at home, rate themselves as speaking English less than “very well.”

SOURCE: U.S. Census Bureau, 2009 American Community Survey 1-year Estimates.

Downloaded on 8/1/2011 from http://factfinder.census.gov/servlet/STTable?_bm=y&-geo_id=01000US&-qr_name=ACS_2009_1YR_G00_S1601&-ds_name=ACS_2009_1YR_G00_&-lang=en&-redoLog=false&-format=&-CONTEXT=st

Often times, in the healthcare setting, staff and providers assume that because they can easily converse with a patient about day-to-day topics in English, described in the education literature as basic interpersonal communicative skills (BICS), the patient does not need an interpreter for healthcare discussions. In many instances health care encounters require cognitive academic language proficiency (CALP). <http://www.wou.edu/~lpearso/website/Cummins.html>. It may take English language learners years to develop CALP, the higher level of proficiency needed to deal with technical information, words, and concepts not part of everyday conversation, such as those typically used in health and mental health care. (Cummins, 1979) Thus, it is vital for healthcare providers and settings to make sure patients understand the information provided to them or questions asked of them.

As such, service providers should implement policies and procedures to provide access to services and information in appropriate languages other than English to ensure that persons with limited English proficiency are effectively informed and effectively participate in any benefit.

The team decides to stop her antibiotics and consults the Nephrology service for a renal biopsy for presumed lupus nephritis. They begin her on Solu-Medrol 60 mg IV every 24 hours empirically. Her subsequent serologies and ANA by immunofluorescence return.

ANA 1:1280, speckled

Anti-Sm +

Anti-RNP +

Anti-ds-DNA + (1202)

Anti-SSA+

The team discusses with the Rheumatology service what her treatment course will consist of in general. Although they agree that she will need prednisone and another agent for her presumed nephritis (to be determined after biopsy results), it is determined that she should also be placed on hydroxychloroquine.

Do you think that exposure to the antibiotic may have triggered her SLE flare?

Flares of SLE often occur without a known inciting event. However, there are several situations that increase the risk of a flare. Medications containing sulfonamides have been reported to trigger the onset of lupus flares in those with pre-existing disease. This is different from drug-induced lupus, which can cause a transient and often milder presentation of SLE that resolves with withdrawal of the inciting medication. Other risks for SLE flare include exposure to sunlight, estrogen (exogenous source, such as birth control pills, or pregnancy), lack of adherence to medications, and severe emotional or physical stress. In this case, the patient most likely had lupus prior to receiving her antibiotic, and the sulfa antibiotic may have contributed to her flare of disease activity.

Review this patient's manifestations of SLE, both clinically and serologically.

The students should identify her nephritis, cognitive impairment, mucocutaneous manifestations, fever, ANA, anti-Smith, anti-RNP, anti-dsDNA, anti-SSA, hypocomplementemia as her predominant manifestations of SLE.

All of these severe manifestations present in a limited English-speaking Mexican female with poor access to care secondary to her place of residence and with family having limited resources to buy medications and navigate the medical system.

How will these socioeconomic circumstances impact her care, treatment, and potentially her outcome?

In Dr. Diamond's lecture, there is a nice description of issues affecting minority groups with systemic lupus erythematosus, specific to patient, physician, and society. These items should be reviewed with the students with emphasis on the fact that data demonstrate worse outcomes, such as renal transplant, in these patient populations.

Poor adherence to medical regimens is often attributed to the patients' lack of insight into their illness. As we saw in this case, linguistic competency, in particular literacy and health literacy, can be a critical barrier. There are often other barriers, including limited access to care secondary to financial, language, or cultural barriers that limit treatment. Students should be encouraged to

Students should begin to appreciate the multi-dimensional components that go into evaluating and treating SLE flares. Particularly with minority patients and those with linguistic and other cultural barriers, the treatment of flares goes beyond just medicinal treatment. Hispanic patients with SLE in the LUMINA study were reported to be significantly more likely to have abrupt onset of their disease. Figure 1 illustrates the differences in abrupt onset among Hispanic, black and white patients. (Alarcon et al., 1999)

Therefore, cultural competency is a frequent and critical issue.

What levels of support are important for this patient?

Students may naturally gravitate to answers related to healthcare access and other social services, which is important and should be discussed. Asking about an SLE patient's supportive network and addressing any concerns about feeling alone or having no help is also an important form of support and component of treatment. Addressing these issues with Hispanic patients may be particularly important. A number of research studies have reported the following:

- Social support (having people in one's life that can provide tangible and emotional help) plays a critical role in the health and well-being of individuals with SLE. (Karlson et al., 1997; Sanchez et al., 2009)
- Poor social support has been found to be associated with higher levels of disease activity, as well as subjective health-related quality of life for SLE patients

