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 multi-system 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.

History of Present Illness

A 38-year-old Spanish-speaking, Mexican female presents with complaints of fatigue, nausea, and vomiting as well as weight loss. Her husband, who speaks little English, provides most of the history.

The emesis is not related to food and consists of yellow fluid and food, but 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 ciprofloxacin, an oral antibiotic from an outside hospital for a presumed urinary tract infection (UTI), although she did not have many symptoms suggestive of UTI. Shortly after the antibiotic was started, she developed abdominal pain and fever. She continued to take the antibiotic for several more days, thinking she would get better with the medication. When symptoms progressed to significant nausea and vomiting, her husband brought her to the university hospital.

Her past medical history is significant for a healthy pregnancy delivered by cesarean section 14 years prior. There is no known family history.

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

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

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

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 patients from diverse groups, such as this Spanish-speaking female. Consider various groups in your community and the potential communication barriers they may face. Consider how your organization, clinic, or hospital could lower these barriers and improve linguistic competence.

She denies using alcohol, tobacco, or drugs, and she lives with her husband and 14-year-old son. All three immigrated to the United States from Mexico and are currently undocumented. Neither she nor her husband graduated from high school. She works cleaning homes, and he works in construction. Together, their income is about $1400 per month. She speaks little English; her husband speaks some English, and her 14-year-old son speaks fluent English.

How might the availability of competent medical interpreters have contributed to this patient's situation? Consider the importance of literacy in good outcomes.

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.[1]

Studies show that persons with low literacy skills are less likely to[2]:

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

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.

Given the lack of translated documents and bilingual staff (or available interpreters), patients who speak only Spanish and patients with limited English proficiency are further challenged by issues of health literacy. 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 level they can understand, using plain language and in a format that maximizes their learning.[3]

Also keep in mind that professional medical interpreters are crucial to obtain an accurate medical history and improve the patient's experience. Relying on family to interpret is not sufficient. Family members are not professional medical interpreters and may be limited by their own literacy issues even for general translation. Also, the patient may not be able to share their medical history and experience freely when a family member is serving as the interpreter.

Physical Exam

Height: 5'7"
Weight: 155 lbs

Vitals: Blood pressure 97/53, heart rate 114/minute, respiration rate 20/minute, O2 saturation 99% on room air, temperature 38.4 °C (101.1 °F)

General: No acute distress

Head, Eye, Ear, Nose, Throat (HEENT): Pale conjunctiva, ulcers noted on the hard palate, no pharyngeal edema

Pulmonary: Clear to auscultation bilaterally

Abdomen: Soft, no rebound tenderness

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 (SLE) are generally not painful and can be found on the hard palate.

Laboratory Data

Sodium: 121 mEq/L*
Potassium: 4.2 mEq/L
Chloride: 98 mEq/L
Bicarbonate: 19 mEq/L*
Blood urea nitrogen: 22 mg/dL
Creatinine: 1.0 mg/dL
Total protein: 5.9 g/dL*
Albumin: 2.0 g/dL*
Calcium: 7.0 mg/dL*
Corrected calcium: 8.6 mg/dL
Total bilirubin: 0.05 mg/dL

Alanine aminotransferase: 63 U/L*

Aspartate aminotransferase: 147*

Alkaline phosphatase: 29 U/L

Lipase: 128 U/L*

Creatine kinase: 275 U/L*

White blood cell count (WBC): 0.824 x 103/μL*

White count differential: 48% segmented neutrophils (absolute neutrophil count 396),* 12% monocytes, 40% lymphocytes (absolute lymphocyte count 330)*

Hemoglobin: 10.5 g/dL*

Hematocrit: 32.2%*

Mean cell volume (MCV): 84 fL

Platelets: 72/μL*

Prothrombin time: 14.0 seconds

Partial thromboplastin time: 48.8 seconds

International normalized ratio: 1.22

Fibrinogen: 231 mg/dL

Urinalysis

3+ protein*

3+ blood*

1+ ketones*

No nitrates

WBC: 11–23/ high power field (hpf)*

Red blood cells (RBC): 2–5/hpf*

Hyaline casts: 2–5/hpf*

Granular casts: 6–10/hpf*

Cellular casts: 2–5/hpf*

Urine pregnancy negative

**Abnormal values*

Abdominal x-ray: No evidence of obstruction

She was admitted to a general medicine team and placed in a room on a regular medical floor. Since she was febrile and met systemic inflammatory response syndrome criteria, the team treated her with antibiotics (vancomycin and piperacillin/tazobactam), while a source infection was investigated. Metoclopramide was ordered, as needed for nausea.

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

Blood cultures: No growth to date

Urine cultures: No growth to date

Hemoglobin: 8.5 g/dL*

MCV: 85 fL

Ultrasound of abdomen was unremarkable

**Abnormal values*

Although her blood and urine cultures were negative, she remained febrile with the same gastrointestinal complaints.

What could be causing her fever at this point?

Consider 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?

Consider 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 the RBC are dysmorphic. Dysmorphic RBC should prompt you 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 (lactate dehydrogenase [LDH], haptoglobin, peripheral smear, Coombs test), iron studies, vitamin B12, and folate levels. They also agree with the connective tissue disease and atypical infectious disease 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 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 RBC in her urine are dysmorphic.

Some of the additional labs ordered return:

C3: 22*
C4: <8*
Erythrocyte sedimentation rate: 65 mm/hr*

LDH: 395 units/L*
Haptoglobin: <30 mg/dL*
Coombs test: +*

Iron: 114 µg/dL, 68%*
Transferrin: 111 mg/dL*

Folate: 7.9 ng/mL
B12: 928 pg/mL

Human immunodeficiency virus (HIV): Negative
Epstein-Barr virus PCR: Negative
Cytomegalovirus PCR: Negative

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

Hepatitis B and C serologies: Negative

**Abnormal values*

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 interpreter, unlike her initial evaluation.

Rheumatology Consult Service Physical Exam

Vitals: Blood pressure 100/70, heart rate 98/minute, respiration rate 14/minute, saturation 99% on room air, temperature: 38.3 °C (100.9 °F)

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

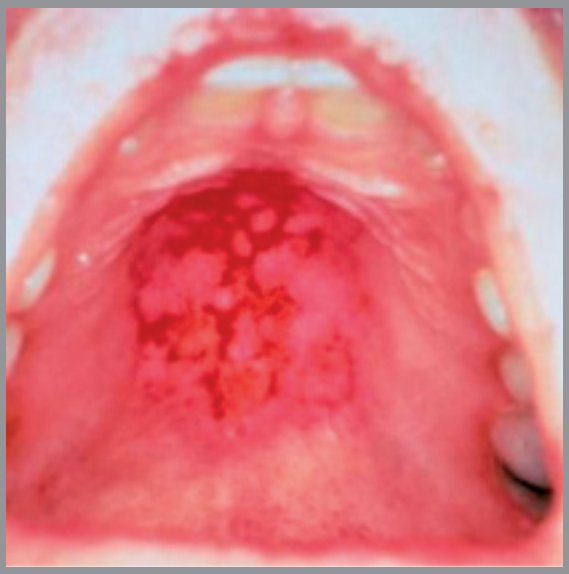
HEENT: pale conjunctiva, normal sclera, angular cheilitis, peeling lips, large deep ulcers on hard palate anterior behind front teeth

Cardiovascular: Normal heart sounds, no murmurs

Pulmonary: Clear to auscultation bilaterally

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

Skin: Fixed livedoid nonblanching lesions on palms (thenar) and fingertips, no digital ulcerations, no nailfold capillary changes



Oral ulcers

Image courtesy of the American College of Rheumatology Image Bank

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 medical interpreter

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 interpreter, which revealed her slightly abnormal mental status. Additionally, her vasculitic rash was identified.

Her antinuclear antibody (ANA) screening test by enzyme-linked immunosorbent assay is positive; however, the immunofluorescence pattern and titer are pending.

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

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

SLE is a clinical diagnosis. Serologic tests alone do not make a diagnosis. The patient must have both the clinical history/physical findings and confirmatory serologic tests.

It is important to have symptoms and findings that can be attributed to SLE. A 38-year-old Hispanic female presenting with months of fatigue, lymphadenopathy, and + ANA is consistent with SLE, but could also be indicative of HIV or other conditions. The important point in diagnosing SLE is one of attribution.

The team decides to stop her antibiotics and consults the nephrology service for a renal biopsy for presumed lupus nephritis. The renal service initiated a workup to determine the cause of the hyponatremia, particularly that it may be contributing to the patient's abnormal mental status. In order to fully evaluate the patient's possible cognitive dysfunction, a neuropsychiatric (NP) evaluation will also need to be obtained. The NP evaluation should be conducted with both cultural and linguistic competence.

She is prescribed intravenous methylprednisolone sodium succinate 60 mg every 24 hours empirically. Her subsequent serologies and ANA by immuno–fluorescence return.

ANA 1:1280, speckled*

Anti-Sm +*

Anti-RNP +*

Anti-dsDNA + (high titer)*

Anti-SSA+*

**Abnormal values*

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

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

In summary, this is a patient who has nephritis, possible cognitive impairment, mucocutaneous manifestations, fever, ANA, anti-Smith, anti-RNP, anti-dsDNA, anti-SSA, and hypocomplementemia as her predominant manifestations of SLE.

All of these severe manifestations present in a Mexican female who speaks little English, has poor access to care, and has limited resources to buy medications and navigate the medical system.

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

Poor adherence to medical regimens is often attributed to the patient's lack of insight into their illness. As this case demonstrates, 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.

It is important 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 medical treatment. Therefore, cultural competency is a frequent and critical issue.

What levels of support are important for this patient?

Consider this patient's access to healthcare and other social services. Asking about an SLE patient's support network and addressing any concerns about feeling alone or having little or 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 who can provide tangible and emotional help) plays a critical role in the health and well-being of individuals with SLE.[4,5]
- 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.
- Over the course of the disease, demographic, behavioral, and psychological variables, including Hispanic ethnicity (sample population from Texas) and social support, are important mediators of disease, while genetic factors do not influence disease activity over time.[6]
- Several studies have reported that Hispanics with SLE have significantly lower levels of reported social support than Whites.[6] One study has demonstrated that an intervention with spouses or other family members and individuals with SLE decreased their reported fatigue, one of this patient's main complaints.[7] Addressing her lack of social support needs to be an integral part of her treatment.

References

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Notes:

Refer to the Lupus Initiative Lecture series for more information.

The images included in this case are for example only and are not those of the individual described in the case.

The production of this activity was supported by Grant Number 7 MPCMP111064-01-00 from the U.S. Department of Health and Human Services Office of Minority Health (HHS, OMH). Its contents are solely the responsibility of the authors and do not necessarily represent the official views of the HHS, OMH.