5-2020

RARE PRESENTATION OF LUPUS: SLE PRESENTING AS SEPTIC SHOCK

Nitesh Gandhi
Henry Ford Health System, NGandhi4@hfhs.org

Yaser Alkhatib
Henry Ford Health System, aalkhat1@hfhs.org

RARE PRESENTATION OF LUPUS: SLE PRESENTING AS SEPTIC SHOCK

Nitesh Gandhi, Ayad H Alkhatib, Department of Internal Medicine

Henry Ford Health System, Detroit, Michigan

Introduction

- Infections are common in patients with SLE because of the immune impairments, but there are only very few cases that present as infection as the initial manifestation of SLE.
- Here we present the case of a patient whose initial presentation was septic shock secondary to Pseudomonas UTI complicated by Pseudomonas bacteremia.

Case Presentation

The patient is an 18-year-old female with no significant past medical history presented with fever, chills, nausea, episodes of vomiting and bilateral lower extremity edema.
Vital signs showed that she is tachycardic to 130s, tachypneic to 30s.
Physical exam showed tense bullae and purpuric plaques bilaterally in lower extremities
Labs were significant for lactic acid of 4.2 and creatinine of 4.54, hemoglobin of 7.1. Infectious workup was done due to concern of septic shock and the patient was found to have UA showing more than 182 WBCs, 114 RBCs, and more than 500 proteinuria.
The patient was intubated, started on broad-spectrum antibiotics and was admitted to MICU for septic shock
Further workup included:
• Urine culture growing Pseudomonas
• Blood cultures growing pan-susceptible Pseudomonas
• BMP showing up rising creatinine
• Autoimmune workup: positive ANA(1:1280) homogeneous pattern, double-stranded DNA antibodies(1:320), SS-A, SS-B, ENA smith/RNP low complement levels, negative C ANCA, p-ANCA, CCP, and anti-GBM antibodies
• Echocardiogram showed moderately sized pericardial effusion is present circumferentially around the entire heart.
• Skin biopsy of tense bullae showed thrombotic vasculopathy with widespread dermal necrosis- most likely represents septic vasculopathy. Skin biopsy tissue culture was positive for Pseudomonas.
• Renal biopsy showed Diffuse Lupus Nephritis, Class 4-G (A).
Management:
• The patient was treated with dialysis(initially SLED and later hemodialysis)
• She also received Cefepime for Pseudomonas infection
• She got Solumedrol 500 mg daily for 3 days and that was transitioned to 60mg daily.
• She was also started on Cellcept given the superiority and better safety compared to Cyclophosphamide for induction therapy in the African-American population, especially with childbearing age(1,2)

Laboratory Findings

Figure 1. Patient's pertinent workup

ANA		Positive
ANA PATTERN		Homogeneous *
ANA TITER 1		> 1:1280
DNA Ab (ds) Crithi...	Positive *	1:320 *
C3 Complement		36
C4 Complement		< 8

Blood Culture (2 Sets)

Gram Stain	Aerobic bottle Gram negative bacilli !
Results	Aerobic bottle Pseudomonas aeruginosa !

Urinalysis

Type		Voided
Volume	mL	12
Color		Amber
Clarity		Cloudy !
Specific Gravity	1.005 - 1.030	1.027
urine pH	5.0 - 7.5	5.0
Protein	mg/dL	>=500 !
Glucose UA	mg/dL	Negative
Ketones	mg/dL	5 !
Bilirubin		Negative
Blood		Large !
Urobilinogen	U/dL	Negative
Nitrite		Negative
Leukocyte Esterase		Large !
RBC	<3 /HPF	114 ^
WBC	<10 /HPF	>182 ^

PATHOLOGICAL DIAGNOSIS:
Left kidney, needle biopsies: Diffuse Lupus Nephritis, ISN/RPS Class 4-G (A).

Discussion

• SLE is a chronic inflammatory multisystem disease with immunological abnormalities and seen more often in women than men.
• In the literature, the infection has been reported as the leading cause of death in a patient with SLE(3) but there are very few cases of SLE which present as infection as the initial manifestation
• There was an almost 5-fold increase in the risk of death from infection in patients with SLE when compared with the general population(4)Factor contributing to this include SLE causing neutropenia, lymphopenia, hypocomplementemia, renal involvement, neuropsychiatric manifestations, and the use of glucocorticoids and other immunosuppressive drugs.

Conclusion

- Physicians should always consider the possibility of complement deficiencies in young adult patients presenting with severe infections and the presence of hypocomplementemia should suggest the possibility of SLE, even in patients with no prior history of SLE and patient should undergo further workup to diagnose Lupus.(5)
- In this situation, the patient should be treated for both infection and SLE.

References

1. N Engl J Med. 2005 Nov 24;353(21):2219-28.
2. Arthritis Care Res (Hoboken). 2012 Jun; 64(6): 797–808.
3. D. B. Hellmann, M. Petri, and Q. Whiting-O'Keefe, "Fatal infections in systemic lupus erythematosus: the role of opportunistic organisms," Medicine, vol. 66, no. 5, pp. 341–348, 1987.
4. M. Yurkovich, K. Vostretsova, W. Chen, and J. A. Aviña-Zubieta, "Overall and cause-specific mortality in patients with systemic lupus erythematosus: a meta-analysis of observational studies," Arthritis Care and Research, vol. 66, no. 4, pp. 608–616, 2014
5. 5M. K. Shaughnessy, D. N. Williams, and B. Segal, "Severe infection with encapsulated bacteria as the initial presentation of systemic lupus erythematosus: two case reports and a review of the literature," JMM Case Reports, 2014.