Rapid Progression to Cardiac Tamponade in Systemic Lupus Erythematosus

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CASE PRESENTATION

Initial Presentation and History

A 24-year-old woman presented to the emergency department with complaints of rash on her face and legs, progressive weakness, and shortness of breath over the previous 3 weeks. She reported rapid weight loss of 25 lb over 3 to 4 weeks and patchy loss of scalp hair. The patient denied chest pain, fever, loss of consciousness, blood in stool, excessive menstrual blood loss, joint pain, or any urinary complaints. The patient had no past medical history.

Physical Examination and Laboratory Evaluation

Physical examination revealed a temperature of 97.2°F (36.2°C), heart rate of 147 bpm, respiratory rate of 20 breaths/min, and blood pressure of 100/65 mm Hg. Head and neck examination revealed conjunctival pallor. Cardiac examination showed an active precordium with loud heart sounds. Skin examination showed hyperpigmented indurated plaques in a malar distribution on the cheeks and across the bridge of the nose with satellite lesions on the chin and forehead; palpable purpura on both lower extremities; and nonscarring patches of alopecia in the scalp.

Significant results from laboratory tests performed on admission are shown in **Table 1**. The patient's high ferritin, normal transferrin and transferrin saturation, and low erythropoietin levels relative to hematocrit suggested anemia of chronic disease. Positive antinuclear antibody (ANA), antibody to double-stranded (ds) DNA, and anti-Smith (Sm) antibody titers suggested systemic lupus erythematosus (SLE). Proteinuria on urinalysis suggested possible renal involvement of SLE. Electrocardiogram showed low-voltage sinus tachycardia. Chest radiograph showed an enlarged cardiac silhouette and "water-bottle heart" suggestive of pericardial effusion (**Figure**). The patient had 5 of the 11 American College of Rheumatology criteria for classifying SLE: malar rash, serositis (pericardial effusion), renal involvement (proteinuria greater than 0.5 g/day), immunologic disorder (positive anti-dsDNA and anti-Sm antibodies), and positive ANA test result (**Table 2**). A diagnosis of SLE was made based on the clinical findings and positive findings on laboratory tests.

Hospital Course

The patient was admitted with disseminated lupus. Evaluation with 2-dimensional echocardiography was performed on admission, which showed moderate pericardial effusion. After admission, the patient had progressively worsening shortness of breath and increasing tachycardia. Distended neck veins were noted on examination 3 days after admission. Repeat echocardiography showed a large pericardial effusion and right ventricular diastolic collapse, suggestive of tamponade physiology, along with right ventricular systolic pressure of 51 mm Hg and a left ventricular ejection fraction of 65%. Pericardiocentesis was done, which yielded 500 mL of straw-colored pericardial fluid; pericardial pressure was 11 mm Hg. Fluid analysis showed a red blood cell count of 9550/μL, a white blood cell count of 90/μL, a protein level of 6.7 g/dL, glucose level of 124 mg/dL, lactate dehydrogenase level of 370 U/L, and an ANA titer of 1:1280. Smears and cultures were negative for infection. Pericardial fluid cytology was negative for malignancy.

A percutaneous kidney biopsy was performed to assess the nephritic-range proteinuria; histologic evaluation of the specimen revealed findings consistent with class II lupus nephritis (mesangial glomerulonephritis). Skin biopsy of her leg showed necrotizing vasculitis of the deep dermal vessels.

The patient was started on prednisone 60 mg daily and hydroxychloroquine 200 mg twice daily and showed rapid improvement. Repeat 2-dimensional echocardiography performed several days after pericardiocentesis did not show reaccumulation of pericardial fluid. Three

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Table 1. Laboratory Values on Day of Admission

Laboratory Tests	Value	Normal
Hemoglobin (g/dL)	5	12.0–15.5
WBC count (per μL)	4.6×10^3	$3.5–10.5 \times 10^3$
Urinalysis		
Protein (mg/d)	> 300	Negative or trace, < 100
WBCs (per hpf)	0–8	—
RBCs (per hpf)	5	0–3
Leukocyte esterase	Negative	—
Nitrites	Negative	—
24-H urine protein (mg/24 h)	2104	77–197
Anemia work-up		
RBC count (per μL)	1.86×10^6	$3.9–5.0 \times 10^6$
Hematocrit (%)	16.5	35–45
MCV (fL)	88.7	82–97
MCH (pg)	26.9	28–34
MCHC (g/dL)	30.3	32–36
Reticulocytes (proportion of RBCs, %)	1.2	0.3–2.0
Reticulocyte count (per μL)	0.02×10^6	$0.02–0.10 \times 10^6$
Serum iron (μg/dL)	37	40–150
Ferritin (ng/mL)	912	10–291
Transferrin (mg/dL)	168	210–370
Transferrin saturation (%)	22	16–50
Haptoglobin (mg/dL)	200	30–200
Folic acid (ng/mL)	12.4	2.5 to 20
Vitamin B_{12} (pg/mL)	747	239–931
Erythropoietin (mU/mL)	86 mU/mL	—
Antibody assays		
Antinuclear antibody titer	1:1280	—
Anti-dsDNA antibody titer	1:1280	—
Anti-Sm antibody (U)	100	0–49
Anti–SS-A antibody (U)	205	0–49
Anti–SS-B antibody (U)	313	0–49
Ribonucleoprotein antibody (U)	76	0–49
SCL-70 antibody (U)	5	0–49
Anticardiolipin IgG (GPL)	37.8	< 15
Immunologic assays		
IgM (MPL)	19.3	< 15
Complement (mg/dL)		
C3	39	70–200
C4	< 2	15–50

ds = double-stranded; hpf = high-power field; MCH = mean corpuscular hemoglobin; MCHC = mean corpuscular hemoglobin concentration; MCV = mean corpuscular volume; RBC = red blood cell; WBC = white blood cell.

Figure. Chest radiograph showing enlarged cardiac silhouette and "water-bottle heart" suggestive of pericardial effusion.

units of packed red blood cells were given to correct the patient's anemia. Twelve days after admission, the patient was discharged on a tapering dose of prednisone, hydroxychloroquine 200 mg twice daily, erythropoietin, and ferrous sulfate and advised to follow-up with her primary care physician and a rheumatologist.

DISCUSSION

SLE is a multi-organ disease that often involves the heart. The cardiac manifestations of SLE include pericarditis, myocarditis, endocarditis, and conduction system abnormalities.[1] Pericarditis is common in late-onset SLE (incidence, 27%),[2] and its clinical,[3–5] anatomic, pathologic, and echocardiographic characteristics have been well described.[6–9] However, this involvement is usually mild pericardial effusion,[3,4,10] with cardiac tamponade developing in only 6% of cases of SLE with pericarditis. Patients with SLE may present with pericardial involvement as the initial manifestation of disease,[3,4] but only a few cases have been reported where cardiac tamponade was the initial manifestation of SLE.[11–15] The low incidence of cardiac tamponade despite the high frequency of pericarditis in SLE may be due in part to the widespread use of steroids and nonsteroidal anti-inflammatory drugs, which are effective in reducing pericardial inflammation.[11]

Cardiac tamponade develops when a critical amount of fluid accumulates in the pericardium, resulting in diminished blood flow to the ventricles.[2] Typical symptoms and signs include dyspnea, orthopnea, chest tightness, jugular venous distention, pulsus paradoxus, hypotension, and distant heart sounds.[12,16] The diagnosis of tamponade is confirmed by echocardiography.[2] Pericardiocentesis and corticosteroids are the initial

Table 2. Revised Criteria for Classification of Systemic Lupus Erythematosus

Criterion	Description
Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
Photosensitivity	Skin rash as a result of reaction to sunlight, by patient history or physician observation
Oral ulcers	Oral or nasopharyngeal ulceration observed by physician
Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
Serositis	Pleuritis *or* pericarditis
Renal disorder	Persistent proteinuria > 0.5 g per day or > 3+ *or* cellular casts (red cell, hemoglobin, granular, tubular, or mixed)
Neurologic disorder	Seizures *or* psychosis
Hematologic disorder	Hemolytic anemia with reticulocytosis, *or* leukopenia < 4000/μL on 2 or more occasions, *or* lymphopenia < 1500/μL on 2 or more occasions, *or* thrombocytopenia < 100,000/μL
Immunologic disorder	Antibody to DNA, *or* antibody to Sm, *or* positive finding of antiphospholipid antibodies based on either an abnormal serum level of IgG or IgM anticardiolipin antibodies, a positive test result for lupus anticoagulant, or a false-positive serologic test for syphilis known to be positive for at least 6 months and confirmed by *Treponema pallidum* immobilization or fluorescent treponemal antibody absorption test
Antinuclear antibody	Abnormal titer

Adapted with permission from Tan EM, Cohen AS, Fries JF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982;25:1271–7; *and* Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1997;40:1725.

treatment of choice.[12,16] Antimalarial drugs have shown added benefits.[17]

In summary, this patient presented with cardiac tamponade and severe anemia as manifestations of SLE. Accurate and timely diagnosis of these rare manifestations of SLE may be lifesaving. **HP**

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