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CASE REPORT

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Lupus and antiphospholipid syndrome diagnosis in a young female presenting with multiple non-specific symptoms

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Abstract

A female in her 30s with no known past medical conditions presented to North Shore University Hospital with three weeks of intermittent fevers, fatigue, and malaise. She also reports generalized body pain, myalgias, ear pain, throat pain, and joint pain. She denies rash. She also reports nausea, vomiting, and diarrhea and a 10-pound weight loss in one week. She had a miscarriage at 16 weeks of gestation. She met systemic inflammatory syndrome criteria on admission as she was febrile to 102.9 Fahrenheit, tachycardic to 120 beats per minute, tachypneic to 22 breaths/minute, and had leukopenia with white blood cell count of 2,470 per cubic milliliter. Labs were significant for pancytopenia, elevated transaminases, low C3 complement level, double stranded DNA > 1000 IU/mL, anti-nuclear factor positive 1:1280, and positive antiphospholipid serology. She was treated with steroids and hydroxychloroquine and was started on warfarin.

Background

Lupus is an autoimmune condition that can affect multiple organ systems. A young female presenting with multiple non-specific and seemingly unrelated symptoms should be evaluated for lupus.

Case presentation

A female in her 30s with no known past medical conditions presented to North Shore University Hospital with 3 weeks of intermittent fevers, fatigue, and malaise. She also reports generalized body pain, myalgias, left ear pain, throat pain, and diffuse joint pain. She denies episodes of morning stiffness, numbness or changes in the color of her distal extremities with cool environments. She denies changes in vision or rash. She also reports nausea, vomiting, and diarrhea over this time period. She denies recent travel or sick contacts. She denies unusual foods or raw foods. She does report a 10-pound weight loss in one week and consistent fevers up to 104 Fahrenheit. On further review, she had a miscarriage at 16 weeks of gestation, between the birth of two healthy children. On admission, she was febrile to 102.9 Fahrenheit, tachycardic to 120 beats/minute, normotensive at 102/59, and tachypneic to 22 breaths/minute but on room air. Her weight was 67.5 kg (kG), height was 165.1 cm, and Body Mass Index (BMI) was 24.8 kG/meter squared. On physical exam, she had polyarthralgia of primarily the large joints including bilateral knees and left shoulder along with intermediate joints such as bilateral ankles, wrists, and elbows without signs of tenosynovitis.

Objective

Labs were significant for pancytopenia (Table 1), mild transaminases elevation (Table 1), mildly elevated urinary Protein/Creatinine ratio 0.33 (Table 2), hypocomplementemia, elevated double stranded deoxyribonucleic acid (dsDNA) > 1000 IU/mL, elevated anti-nuclear factor (ANA) 1:1280, beta 2 glycoprotein antibody screen

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Table 1 Table including a complete blood count and comprehensive metabolic panel

Complete Blood Count	**Reference Ranges:**
White Blood Cells: 2,470/uL,	[3800–1050/uL]
Hemoglobin 10.2 g/dL	[11.5–15.5 g/dL]
Platelet 104,000/uL	[150,000–400,000/uL]
Comprehensive Metabolic Panel	**Reference Ranges:**
Na + 133 mmol/L	[135–145 mmol/L]
K + 3.9 mmol/L	[3.5–5.3 mmol/L]
Blood Urea Nitrogen 10 mg/dL	[7–23 mg/dL]
Creatinine .73 mg/dL	[0.5–1.30 mg/dL]
Total Bilirubin 0.6 mg/dL	[0.2–1.2 mg/dL]
Aspartate aminotransferase: 62 U/L	[10–40 U/L]
Alanine aminotransferase of 48 U/L	[10–45 U/L]
Alkaline Phosphatase of 46 U/L	[40–120 U/L]
Serum albumin 3.3 g/dL	[3.3–5.0 g/dL]

Table 2 Urinalysis showing a Urinary protein/creatinine ratio of 0.33

Urinalysis	**Reference Range**
Urinary protein 19 mg/dL	[0–12 mg/dL]
Urinary creatinine 56 mg/dL	[Reference range not established given variability]
Protein/Creatinine Ratio 0.33	[0–0.2 Ratio]

positive, Anticardiolipin Immunoglobulin M (IgM) positive, Phosphatidylserine/Prothrombin IgM positive, and anti-Sjogren's Syndrome related antigen (anti-SSA) antibody positive (Table 3). Blood and urine cultures were negative.

Computed Tomography (CT) of the chest/abdomen as well as Magnetic Resonance (MR) imaging of the head with intravenous contrast showed no acute findings.

Treatment

She was initially started on broad spectrum antibiotics with vancomycin and piperacillin-tazobactam which were discontinued when blood and urine cultures were negative. She was treated with steroids (solumedrol 40 mg daily) along with hydroxychloroquine and had relief of some symptoms.She was also started on warfarin given the positive serology for antiphospholipid syndrome. We arranged rheumatology follow up outpatient.

Discussion

Lupus is a chronic autoimmune inflammatory condition that can affect multiple systems. The four types include Systemic Lupus Erythematous, drug-induced lupus, neonatal lupus, and cutaneous lupus.

Based on the 2019 EULAR/ACR classification system, our patient met criteria for a diagnosis of lupus. The entry criterion was positive as antinuclear antibodies (ANA) was greater than 1:80. For the additive criteria, she got 2 weighted points for fever, 6 points for polyarthralgia, 2 points for presence of lupus anticoagulant, 3 points for low C3 complement level and 6 points for being positive for anti-dsDNA antibody. This gives her a weighted score of 19 which is greater than 10 required for lupus classification [1].

Hydroxychloroquine is an anti-malarial drug that is recommended in many patients with lupus and can relieve symptoms as well as prevent flares [2]. Given the risk of retinopathy, it is necessary to have regular ophthalmological examinations while on the medication. Glucocorticoids are effective in the management

Table 3 Table showing other laboratory workup including complement, infectious, and autoimmune antibody workup

Complement, Infectious, and Autoimmune antibody workup	**Reference Range**
Complement component 3 (C3) of 60 mg/dL	[81–157 mg/dL]
Complement component 4 (C4) of 16 mg/dL	[13–39 mg/dL]
Procalcitonin of 0.12 ng/mL	[.02-.10 ng/mL]
Anti-nuclear factor (ANA) positive 1:1280	[< 1:80]
double stranded deoxyribonucleic acid (dsDNA) > 1000 IU/mL	[< 29 IU/mL]
beta 2 glycoprotein antibody screen positive	[Negative]
Anticardiolipin Immunoglobulin M (IgM) positive	[Negative]
Phosphatidylserine/Prothrombin IgM positive	[Negative]
anti-Sjogren's Syndrome related antigen (anti-SSA) antibody positive	[Negative]
Treponema Pallidum antibody negative	[Negative]
Human Immunodeficiency Virus (HIV) negative	[Negative]
Chlamydia and Gonorrhea Polymerase Chain Reaction Amplification negative	[Negative]
Acetylcholine receptor antibody negative	[Negative]

of serious lupus complications such as lupus nephritis, cerebritis, pleuritis, or pericarditis. Steroids can also provide rapid symptom relief, but the long-term goal should be to minimize daily dosage to 7.5 mg prednisone or less due to side effects [2]. To this aim, early initiation of immunosuppressive agents can facilitate the tapering of steroids. Methotrexate and azathioprine are common immunosuppressive agents. Mycophenolate mofetil is particularly useful in renal and non-renal disease, but not in neuropsychiatric complications. Cyclophosphamide can be used in organ-threatening disease and refractory cases, though it has significant gonadotoxic effects [2]. The biologic belimumab is approved for lupus treatment in adult and children, with other biologics (anifrolumab, ustekinumab) in clinical trials [3]. Biologics are particularly useful in extrarenal disease with inadequate control on first line treatments. Lastly, Voclosporin is approved in the treatment of lupus nephritis [4].

Antiphospholipid (APS) syndrome is also common in patients with lupus and increases risk for thrombotic and obstetric complications. Low dose aspirin and hydroxychloroquine may be used as part of preventative treatment. Once a patient has an arterial or venous thrombotic event, anticoagulation with warfarin is recommended. In a recent trial of 120 patients with lupus-APS syndrome, patients randomized into a Rivaroxaban group had increased thrombotic events (19%) compared to Warfarin (3%), and the trial was terminated early. Thus, the use of novel oral anticoagulants in the treatment of antiphospholipid syndrome should be avoided, and warfarin is the preferred agent [5]. In pregnancy, warfarin may be substituted with heparin.

Lupus is a multisystem disease that can present with multiple non-specific complaints, and early diagnosis and management is essential.

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Availability of data and materials
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Declarations

Ethics approval and consent to participate
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Consent for publication
Written Informed consent was obtained from the patient included in this report.

Competing interests
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