Purpura fulminans: A rare presentation of antiphospholipid syndrome

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ABSTRACT

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CASE REPORT

A 72-year-old male with no significant past medical history who was admitted with worsening bilateral lower extremity discoloration and swelling for three days. He complained of intermittent chest pain and generalized fatigue, but denied dyspnea, fever, weight loss, trauma or any episodes of bleeding. Physical examination was remarkable for non-blanching and non-tender erythematous rash with branched configuration extending up to the knees associated with cold and cyanotic toes but palpable distal pulses bilaterally. There was 1+ bilateral pitting edema extending up to the knees. Investigations (Table 1) were significant for neutrophilic leukocytosis, normocytic anemia and thrombocytopenia. Coagulation profile was unremarkable. D-dimer and fibrinogen levels were noted to be elevated. Renal and hepatic functions were within normal limits and there were no electrolytes or metabolic derangements. Deep vein thrombosis (DVT) studies and computed tomography (CT) pulmonary angiography failed to show large vessel thromboembolic phenomena. Immunology testing revealed positive RF and mildly reduced C4. Human immunodeficiency virus (HIV), mycoplasma, anti-nuclear antibody (ANA), C3, and anti-neutrophil

cytoplasmic antibody (ANCA) serologies were negative. Ankle brachial index and peripheral vascular resistance were normal. Patient was started on prednisone 1 mg/kg and broad spectrum antibiotics on the first day of hospitalization to cover for septic versus immunologic phenomena. Blood cultures were negative and antibiotics were discontinued accordingly. Patient's condition was deteriorating evident by worsening skin necrosis and formation of widespread hemorrhagic blisters and ecchymosis (Figures 1 and 2). Platelet count further dropped. Patient remained hemodynamically stable during the course; however, he was transferred to medical ICU for close monitoring. Intravenous immunoglobulins (IVIG) and heparin infusion were initiated, and he was switched to high dose methylprednisolone intravenously. Serology testing revealed elevated phosphatidylserine IgA, IgM and IgG, and anticardiolipin (aCL) IgM levels. Serology testing for cryoglobulinemia and systemic lupus erythematosus (SLE) was negative. The diagnosis of idiopathic antiphospholipid syndrome was made based on the clinical picture and the positive serology. Heparin infusion was started. During the hospital course, patient's symptoms significantly improved. Platelet count trended up and skin necrotic changes started to resolve. Intravenous immunoglobulins and intravenous steroids were discontinued. The patient was started on warfarin and discharged to nursing home after a total duration of two weeks in hospital.

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DISCUSSION

Antiphospholipid syndrome is an autoimmune multisystem disorder characterized by arterial, venous, or small vessel thromboembolic events and/or pregnancy complications in the presence of persistent antiphospholipid antibodies (aPLs) [1]. Presenting symptoms typically include blood clots, stroke, peripheral arterial thrombosis, or repeat miscarriages [2]. The development of life-threatening acute retiform and widespread purpuric lesions (purpura fulminans) at the time of presentation has been rarely reported [3, 4]. The mainstay of treatment for antiphospholipid syndrome

Table 1: Lab results

Test	Result	Reference Range
White blood cell count	13.1x10 ³ /μL	4.8–10.8x10 ³ /μL
Hemoglobin	12.6 g/dL	14.0–18.0 g/dL
HCT	38.3 %	42–52 %
MCV	85.7 fL	80–94 fL
MPV	10.9 fL	7.2–10.4 fL
Platelet	65 K/μL	130–400x10 ³ /μL
Neutrophils Auto.	87 %	44–80%
Lymphocytes Auto.	3.4 %	13–43 %
Monocytes Auto.	9.3 %	2.0–15 %
Eosinophils Auto.	0.1 %	0.0–3.0 %
Basophils Auto.	0.2 %	0.0–3.0 %
PT	26 secs	10.1–13.0 secs
PTT	26 secs	26.6–34.2 secs
INR	1.2	0.9–1.1
D-dimer	6500 ng/mL	45–500 ng/mL
Fibrinogen	930 mg/dL	230–458 mg/dL
BUN	30 mg/dL	9–20 mg/dL
Creatinine	0.7 mg/dL	0.7–1.3 mg/dL
Sodium	136 mEq/L	137–145 mEq/L
Potassium	3.8 mEq/L	3.5–5.1 mEq/L
CO ₂	26 mEq/L	22–30 mEq/L
Troponin	< 0.012 ng/mL	< 0.034 ng/mL
Creatine Kinase (CPK)	23 U/L	55–170 U/L
Rheumatoid Factor	80 IU/mL	< 15 IU/mL
C4	13.5 mg/dL	16–48 mg/dL
Anticardiolipin IgM	15 MPL	< 12 MPL
Phosphatidylserine IgM	49 U/mL	< 25 U/mL
Phosphatidylserine IgG	24 U/mL	< 10 U/mL
Phosphatidylserine IgA	45 U/mL	< 20 U/mL

Figure 2: Ecchymotic changes noted on the planter surface of right foot.

includes the following antithrombotic medication: heparin, warfarin and aspirin [5–8].

CONCLUSION

Antiphospholipid syndrome is a rare but potentially life threatening disease (especially catastrophic antiphospholipid syndrome). It should be suspected in patients with unexplained skin necrosis and thrombocytopenia, and those presenting with purpura fulminans, when no apparent etiology can be found. Prompt treatment with steroids, intravenous immunoglobulins and anticoagulation can be lifesaving.

Keywords: Anticoagulation, Antiphospholipid syndrome, Purpura fulminans, Thrombocytopenia

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Figure 1: (A) Necrotic, hemorrhagic and tense bullae noted on the dorsum of the right foot, and (B) Necrotic, hemorrhagic and tense bullae noted on the dorsum of the right foot.

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Author Contributions

Ahmed S. Mahmood – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Noor Q. Omar – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sudheer Chauhan – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Jose Cervantes – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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