Purpura Fulminans: A Rare Presentation of Antiphospholipid Syndrome

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Introduction

Antiphospholipid syndrome (APS) 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). Presenting symptoms typically include blood clots, stroke, peripheral arterial thrombosis, or repeat miscarriages. Scarce literature exists about the development of life-threatening acute retiform and widespread purpuric lesions (purpura fulminans) at the time of presentation.

Clinical Case

We present a case of 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 or any episodes of bleeding. Physical examination was remarkable for non-blanching, erythematous rash with branched configuration extending up to the knees, with cold and cyanotic toes but palpable distal pulses bilaterally. Lab work was significant for neutrophilic leukocytosis (13.1K/µL), hemoglobin=12.6 g/dl and platelet count=65K/µL. Coagulation profile showed: PT=26 seconds, INR=1.2, PTT=26 seconds, D-dimer=6500 ng/ml and fibrinogen=930 mg/dl. Comprehensive metabolic panel and serial troponins were unremarkable. DVT studies and CT pulmonary angiography failed to show large vessel thromboembolic phenomena. Immunology testing revealed positive rheumatoid factor (80 IU/ml) and mildly reduced complement C4 (13.5 mg/dL). Hepatitis profile, HIV, mycoplasma, ANA, C3, and ANCA serologies were negative. Ankle brachial index and peripheral vascular resistance were normal. Patient was started on empiric steroids and antibiotics on the first day of hospitalization. There was no bacterial growth on blood cultures and antibiotics were discontinued. Initially, there was a poor response to steroids. Skin necrosis worsened with formation of widespread hemorrhagic blisters and ecchymosis along with a further drop in platelet count. 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 along with an increment of steroid dose. Serology testing revealed elevated phosphatidylserine IgA, IgM and IgG, and anticardiolipin (aCL) IgM levels. Cryoglobulins and aCL IgG were negative. The diagnosis of idiopathic APS was made based on the clinical picture and the positive serology. During the hospital course, patient's symptoms significantly improved. Platelet count trended up and skin necrotic changes started to resolve. The patient was started on warfarin and was discharged to nursing home after two weeks hospital stay..

Discussion

APS is a rare but potentially life threatening disease (especially catastrophic APS). 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, IVIG and anticoagulation can be life saving..

Figure 1.

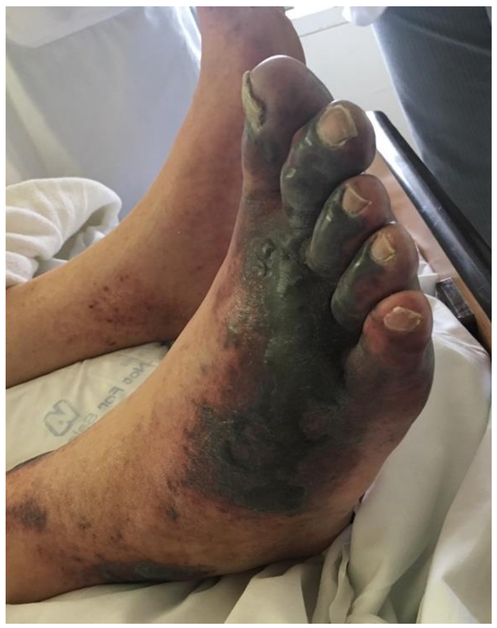

Figure 1: Necrotic, hemorrhagic and tense bullae were noted on the dorsum of the right foot.

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