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Catastrophic antiphospholipid syndrome and sarcoidosis: A case report

Nova Thomas John¹, Madhavi Gorusu¹, Katrina Collins² and Joseph Di Giuseppe²¹Starling Physicians, USA²Hartford Hospital, USA

Catastrophic antiphospholipid syndrome (CAPS) is a systemic autoimmune disease which occurs in <1% of patients with Antiphospholipid syndrome (APS). It is the most severe variant of the classic APS, characterized by histopathologic and clinical evidence of widespread small vessel microthrombi. The resulting inflammatory cytokine storm causes multi-organ failure over a short period and laboratory confirmation of high antiphospholipid antibody titers. Sarcoidosis is a systemic inflammatory disorder characterized by granulomatous inflammation of various organs. Although the association of APS and sarcoidosis may be explained by shared immune dysregulation, cases with concurrent sarcoidosis and APS are extremely rare. Here, we present the 12th reported case, presenting with digital gangrene and review the literature on CAPS. A 66 year-old gentleman presented with rapidly progressive ischemic changes of extremities with skin ulcerations and gangrene of peripheral digits. Autoantibodies testing revealed elevated levels of anti-beta2 glycoprotein IgM and anti-cardiolipin IgM antibodies. Skin ulceration biopsy showed vasculitis with intravascular microthrombi deposition. First-line treatment was initiated for "Probable CAPS" with anticoagulation, glucocorticoids and therapeutic plasma exchange. Subsequent, bone marrow biopsy workup for acute leucopenia with lymphopenia, revealed non-necrotizing granuloma, suggestive of sarcoidosis. This was further substantiated with high serum Angiotensin converting enzyme level. CAPS is a challenging systemic disease requiring a high index of clinical awareness, as outcomes are poor without prompt recognition and early initiation of targeted multimodal therapy. This case highlights the need for a collaborative team approach. It is also the first case reported of probable CAPS associated with sarcoidosis of bone marrow.

Biography

Nova Thomas John has completed her MBBS degree from Kasturba Medical College, Manipal University, Karnataka, India and Internal Medicine Residency from University of Illinois Urbana-Champaign, Illinois, USA. She is currently practicing as a Hospitalist Medicine Physician with Starling Physicians Group at Hartford Hospital, Connecticut, USA since August 2015.

Nova.John@hhchealth.org

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