

ANTIPHOSPHOLIPID ANTIBODY SYNDROME

A1 Bleeding in a Patient with Lupus Anti-Coagulant: A Case Report

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We report the rare simultaneous occurrence of Anti-Phospholipid Antibody Syndrome (APAS) and Acquired Factor VIII Inhibitor in a 49-year old female who presented with acute deep vein thrombosis of the lower extremities and bleeding diathesis. Both conditions were brought about by autoimmune mechanisms directed at components of the coagulation cascade but caused opposing shifts in the hemostatic balance. Steroid administration and intravenous immunoglobulin proved beneficial in the management of this patient.

CLINICAL PRESENTATION. A 49-year old previously healthy Filipino female is admitted for acute bilateral leg swelling following a 3-month course of spontaneous hematoma formation and one episode of spontaneous hematuria.

PHYSICAL EXAMINATION. On admission, the patient is pale with multiple cutaneous hematomas in the right forearm, posterior aspects of both lower extremities, and the right ankle. Both lower extremities are swollen and a positive Homan's sign is elicited on palpation of the gastrocnemius.

LABORATORY WORK-UP. Lupus anticoagulants were detected in the laboratory manifesting as prolonged aPTT that failed to correct at 1:1 mixing with normal plasma and a prolonged KCT. ANA was positive although blood counts and urinalysis did not show hematologic and renal manifestation compatible with a connective tissue disease, particularly SLE. Likewise, immunofluorescence for anti-dsDNA was negative. Anti-cardiolipin antibodies were not detected.

DIAGNOSIS AND TREATMENT. The diagnosis of APAS was made based on demonstration of lupus anti-coagulant activity following a thrombotic event. Methylprednisolone pulse therapy (MPPT) was given and resolution of the lower extremity swelling was observed thereafter. However, the patient continued to develop spontaneous soft tissue hematomas and hemarthroses of the left hip. Further evaluation for a concomitant coagulopathy showed that Factor VIII activity was < 10% of normal. This was later confirmed to be due to the presence of Factor VIII inhibitor. Intravenous immunoglobulin (IVIG) was then ordered.

OUTCOME. Following the administration of IVIG, no new hematomas were observed and repeat coagulation assays improved. The patient was subsequently discharged. And on out-patient follow-up, all hematomas resolved and coagulation test normalized.

SIGNIFICANCE. Bleeding in a patient with APAS is a rare occurrence. Previous reports have attributed this to manifestations also found in this hypercoagulable state such as thrombocytopenia and platelet function defects. Inhibitors directed towards Factor VIII, X and II have also been reported. Among these, anti-Factor VIII autoantibodies are the most common. Quantitative inhibitor measurement assays are necessary to confirm their presence.

A2 Antiphospholipid Antibody Syndrome in a Patient with Dermatomyositis a Case Report

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OBJECTIVE: This case report aims to describe the clinical course and treatment of a rare case of a patient with an overlap of dermatomyositis and antiphospholipid antibody syndrome.

SUMMARY: This is a case of a 29 year old female who initially presented with a history of fever, proximal muscle weakness, and appearance of Gottrons papules and rashes on the face, trunk and

extremities. Pertinent work-ups included a positive antinuclear antibody, anti-Jo1 and anti-RNP antibodies. CK-MM level was elevated and muscle biopsy was consistent with an inflammatory myopathy. Patient was then diagnosed to have dermatomyositis and was treated with corticosteroids. Six years after the diagnosis, she had onset of progressive dyspnea and Raynaud's phenomenon. Work-up done revealed moderate pulmonary hypertension on 2D-echo and pulmonary scintigraphy showed findings consistent with pulmonary thromboembolism. Activated partial thromboplastin time was prolonged and did not correct with mixing. Kaolin clotting-time test and dilute Russell's viper venom time were also both prolonged. The patient was treated with Enoxaparin and later shifted to high-intensity coumadin. A vasodilator and a diuretic together with steroids were also given. She had regular follow-up with a local physician with an INR ranging from 3-4. Despite this, patient had recurrence of dyspnea and eventually expired.

CONCLUSION: The course of a patient with the novel overlap of dermatomyositis and antiphospholipid antibody syndrome was presented. This is the second case in literature of the concurrence of dermatomyositis with APAS. She was treated with corticosteroids and other immunosuppressive drugs for recurrent flares of dermatomyositis however the continuous state of hypercoagulability secondary to the lupus anticoagulants resulted in the patient demise despite treatment with coumadin. Future studies may provide which therapy is best for this overlap syndrome and determine the definite pathogenesis and prognosis in patient with this association.

A3 Catastrophic Antiphospholipid Antibody Syndrome in an Elderly Male with Systemic Lupus Erythematosus: a Case Report

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Rheumatology Section, Department of Medicine UP-PGH, 2001

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We describe a 71-year-old Filipino male with SLE of 4 years duration who developed intractable thrombocytopenia and acute myocardial infarction. Work-ups revealed elevated CK-MB (140 u/L) and positive troponin I (>150 ng/ml), prolonged KCT (110 sec) and DRVVT (52 sec) and elevated anticardiolipin IgM (18.71 MPL units/ml, NV≤12.5 MPL units/ml) and IgG (23.76 GPL units/ml, NV≤15 GPL units/ml). Patient was diagnosed to have catastrophic antiphospholipid antibody syndrome (APS). Despite therapy with pulse methylprednisolone and pulse cyclophosphamide, patient's medical condition deteriorated and eventually expired due to bleeding secondary to an intractable thrombocytopenia. First coined by Asherson et al in 1992, catastrophic antiphospholipid antibody syndrome is a rare accelerated form of APS characterized by multi-organ thrombotic microangiopathy. Since its description, only 50 patients have been described in the literature mostly occurring in young adult females with primary APS. We are reporting the first case of a catastrophic APS in an elderly male patient with SLE. Catastrophic APS is a potentially life-threatening condition. At present, no therapeutic regimen has proven to be consistently successful hence prognosis has remained poor. The aim of this case report is to increase clinical awareness to the syndrome, which in turn can spur further interest into understanding the pathophysiology of the disease and therefore improve on present management. It is therefore recommended that further research be done in further elucidating its mechanism of action and in improving the therapeutic management.

A4 Antiphospholipid Antibody Syndrome in a Patient with Systemic Sclerosis: a Case Report

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OBJECTIVES: This case report aims to discuss the clinical course of a patient with diffuse systemic sclerosis (SSc) with severe pulmonary hypertension and secondary antiphospholipid antibody syndrome (APAS), and to discuss treatment options for a such a case.

SUMMARY: We describe the third case in literature of APAS in a patient with SSc. This is a 40-year-old female who initially presented with a three-year history of Raynaud's phenomenon and digital pitting and a year and a half later developed progressive diffuse skin thickening. She later had a chronic progressive course of thromboembolism, primarily in the lungs, later diagnosed to be due to APAS. The patient was treated for the scleroderma and was anti coagulated for the APAS but still died, underlying the difficulty in diagnosing and treating such rare cases.