CASE REPORT

Antiphospholipid Antibody Syndrome Presenting with Unilateral Adrenal Hemorrhage
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ABSTRACT
The antiphospholipid antibody syndrome presents with vascular thrombosis which involve both arterial and venous systems. The clinical presentation of antiphospholipid antibody syndrome includes obstetric complications leading to recurrent abortions, presence of circulating antibodies against phospholipids, and multi-organ thromboembolisms. We report a case of a patient who presented with unilateral adrenal hemorrhage and subsequently found to have antiphospholipid antibody syndrome and lupus nephritis.

Key Words: Antiphospholipid antibody syndrome. Recurrent abortions. Unilateral adrenal hemorrhage. Lupus nephritis.

INTRODUCTION
The antiphospholipid antibody syndrome (APS) presents with vascular thrombosis which involve both arterial and venous systems. The clinical presentation of antiphospholipid antibody syndrome includes obstetric complications leading to recurrent abortions, presence of circulating antibodies against phospholipids, and multi-organ thromboembolisms. Antiphospholipid antibodies are also found in 1 - 5% of the healthy adults.1 These are much more prevalent among patients with systemic lupus erythematosis (SLE). The most commonly detected subgroups of antiphospholipid antibodies are lupus anticoagulant antibodies, anticardiolipin antibodies, and anti-b2-glycoprotein-I antibodies.1

The most common manifestation is deep venous thrombosis of the lower limbs, followed by stroke, superficial thrombophlebitis, pulmonary embolism, and transient ischemic attacks. Other infrequent presentations include migraine, renal vein/artery thrombosis, cutaneous ulcers, pulmonary hypertension, cardiac-valve vegetations, and adrenal insufficiency.1,2

Adrenal gland ischemia, associated with the antiphospholipid antibody syndrome, has been reported in the literature less frequently. Most of these cases presented as bilateral adrenal hemorrhage or infarction.3 However; unilateral adrenal hemorrhage in patients with the antiphospholipid antibody syndrome is rare. In a review of 86 patients, only 7 patients had unilateral adrenal hemorrhage.3

Here, we report a case of a patient who presented with unilateral adrenal hemorrhage and was subsequently found to have antiphospholipid antibody syndrome and lupus nephritis.

CASE REPORT
A 30-year married female from Kashmir presented with right sided loin pain which was sudden in onset, continuous, moderate in intensity with no aggravating and relieving factors. She denied any history of vomiting or loose motions. Past medical and surgical history was not significant. No history of fever, urinary problem, trauma, rash, joint pains and hair loss. She went to a local hospital where analgesics were given and pain was relieved temporarily. Drug history was unremarkable. Menstrual history was normal and obstetric history revealed 5 first-trimester abortions.

On general physical examination, a young lady lying on bed, her pulse was 82/minute and blood pressure 130/80 mmHg. She had pallor. Her baseline laboratory tests showed anemia with normal MCV, leukocyte count, and platelets. Biochemistry was showing serum creatinine of 2.27 mg/dl. LFTs were normal. APTT was prolonged (patient = 60 seconds, control = 33 seconds) with normal PT. Urine examination showed active sediment with significant proteinuria. Her ANA and anti-dsDNA was positive and she had hypocomplementemia. Anticardiolipin antibodies were positive. Kidneys were of normal size bilaterally. Her adrenal function tests (serum cortisol and synectin test) were normal.

During her stay in hospital, ultrasound abdomen showed a suprarenal mass. CT scan of abdomen showed right adrenal hemorrhage (Figure 1). A diagnosis of secondary antiphospholipid antibody syndrome with active lupus nephritis and unilateral adrenal hemorrhage was made.

She was managed conservatively with supportive measures like blood products transfusions to prevent
further adrenal hemorrhage. She did not require steroids as her blood pressure remained stable. Later, she was discharged home in stable condition on aspirin and with a plan to perform renal biopsy for classification and management of lupus nephritis.

**DISCUSSION**

Primary adrenal insufficiency or Addison disease is the result of destruction of the adrenal cortex by disease processes like tuberculous adrenalitis or autoimmune diseases. Symptoms and signs of adrenal insufficiency appear after extensive or bilateral involvement. The relationship of primary adrenal failure and the presence of elevated antiphospholipid antibody levels is well documented.3

This patient had secondary antiphospholipid antibody syndrome as suggested by the history of recurrent abortions, prolonged APTT, and positive anticardiolipin antibodies. She also had systemic lupus with nephritis as suggested by positive antinuclear antibodies and active urine sediment along with low complements. She had unilateral involvement of adrenal gland and thus had preserved adrenal function.

The pathogenesis of adrenal insufficiency in APS is not clearly understood. It is not related to the presence of autoantibody and is essentially vascular. The vascular anatomy of adrenal gland is unique in the sense that it has multiple arteries but with a single venous drainage, which predispose patients to thrombosis. The capillaries form a vascular plexus around the zona reticularis. The transition from artery to capillary plexus is abrupt and results in formation of a vascular ‘dam’. The capillary plexus ends in medullary sinusoids that eventually form the central vein. In addition, the structure of this single central vein is unique. The musculature of the vein is not concentric but is composed of thick longitudinal muscle bundles. Fox suggested that this eccentric muscular arrangement of the adrenal veins made them especially vulnerable to the formation of platelet thrombi in pockets of turbulence and local stasis which developed due to bundle contraction. He concluded that the primary event was thrombosis leading to secondary hemorrhage.6 Histopathology results of adrenal glands showed hemorrhagic infarction with vessel thrombosis as the main finding. In such cases, acute adrenal hemorrhage usually occurs after the patient either undergoes surgery or receives anticoagulant therapy for recurrent thromboembolisms.7-9 This patient had no history of use of heparin or recent surgery.

The symptoms of adrenal insufficiency may be associated with extensive or bilateral involvement of adrenal glands. It presents with abdominal pain, hypotension, fever, nausea or vomiting, and weakness. Adrenal insufficiency is more common in primary rather than secondary antiphospholipid antibody syndrome.

In case of antiphospholipid antibody syndrome, a high index of clinical suspicion is necessary for the diagnosis of primary adrenal insufficiency. It is, therefore, mandatory in any antiphospholipid antibody positive patient, who complains of abdominal pain and undue weakness or asthenia, to carry out an initial screening for adrenal hemorrhage and hypoadrenalism. Computed tomography and magnetic resonance imaging are considered the best modalities for the diagnosis as CT scan shows enlarged adrenal gland or distortion by a round or oval mass.

Espinosa and colleagues reviewed 80 published case reports and 6 cases of their own on primary adrenal insufficiency due to antiphospholipid antibody syndrome.10 Of the 86 cases (62.8%) having adrenal gland hemorrhaging or hemorrhagic infarctions, only 5 (5.8%) of the patients with adrenal insufficiency had radiologically documented unilateral hemorrhage.10

So in conclusion, this case is a rare presentation of possibly secondary ASA with adrenal hemorrhage and lupus nephritis.

**REFERENCES**


