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An Atypical Presentation of Antiphospholipid Syndrome

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ABSTRACT

Arterial thrombosis is a major cause of morbidity and mortality both in the United States and worldwide. Although the rate of strokes has fallen by a third each year, about 690,000 people experience a new or recurrent ischemic cerebral vascular accident (CVA). It is estimated that 1 in every 20 deaths in the United States of America is due to a CVA. There are various factors that increase the risk of developing arterial thrombosis. Classically, hypertension, elevated levels of low-density lipoprotein-cholesterol (LDL) and smoking are well-documented cardiovascular dependent risk factors implicated in thrombosis. Furthermore, diabetes, pregnancy, age, chemotherapeutics and infectious burden may also contribute to risk of arterial thrombosis. However, an often overlooked cause of thrombosis include autoimmune syndromes such as Systemic lupus erythematosus (SLE), rheumatoid arthritis, and antiphospholipid syndrome (APS). Specifically, antiphospholipid syndrome is a widely recognized autoimmune prothrombotic risk for both arterial and venous thrombosis. Female patients often present with a history of stillbirth as well as loss of multiple pregnancies which can increase the risk of ischemic stroke and MI. Nevertheless, men can have an atypical presentation of antiphospholipid syndrome. In this case report we are presenting a 31-year-old male with past medical history significant for white coat hypertension and obesity who was brought to the emergency department for evaluation of left arm weakness and sudden onset left facial droop.

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Introduction

Antiphospholipid syndrome (APS) is a clinical autoimmune syndrome characterized by symptoms of arterial and/or venous thrombosis and/or pregnancy morbidity in the presence of antiphospholipid antibodies (aPL) and possible life-threatening multi organ failure [1]. Half of cases in APS are primary, while the other half is associated with other systemic autoimmune diseases [2]. Thromboses are a hallmark of APS, with venous thrombosis being more common than arterial [2]. APS should be considered with occurrence of one or more unexplained arterial or venous thrombotic events, especially in young people under the age of 50. In addition, one or more specific adverse outcomes related to pregnancy in females with APS includes fetal death after 10 weeks gestation, premature birth due to severe preeclampsia, placental insufficiency or multiple embryonic losses (<10 weeks gestation) [1]. While there are a multitude of symptoms, there are no pathognomonic physical exam findings in APS. Nevertheless, abnormal features may be found on examination of the patient such as ischemia of the skin, viscera, and/or central nervous system [1]. Therefore, the diagnosis of APS is based on both clinical criteria and evidence of persistent aPL. While sensitive for APS, transient aPL in the blood is nonspecific and can also be seen in certain infections, medications and malignancies. Some of these infections include syphilis, Lyme disease, Leprosy, hepatitis A, B and C and Epstein-Barr virus, and medications include but are not limited to quinine and amoxicillin [1]. Transient aPL in isolation rarely causes thrombosis [1].

Case presentation

This is a 31-year-old male with a past medical history significant for whitecoat hypertension and obesity who presented with acute onset left facial droop and left arm weakness. The symptoms presented a few hours prior to arriving in the emergency department. The patient reported recent travel from Hawaii to New Jersey. During a layover in Seattle airport, he noted sudden loss of strength and numbness/tingling of his face and left upper extremity. On further questioning, the patient reported a history of a similar episode while on vacation in Hawaii. During the previous episode, the patient mentioned a similar episode of left sided mouth drop which prompted him to go to the nearest emergency room. Computed Tomography (CT) without contrast revealed no acute findings, and the patient's symptoms had resolved prior to his encounter with the physician. He was then discharged home with instructions to follow-up with his primary care physician.

However, his symptoms had returned when he arrived at home which prompted him to once again go to the emergency department.

Upon arrival in the ED he was found to be hypertensive (BP: 166/105 mmHg). On neurological examination, the patient was alert and oriented to time, place, and person. Patient presented with a right facial droop insinuating a defect in cranial nerve VII along with left arm pronator drift. Motor strength was 5/5 bilaterally in all four extremities with sensation grossly intact. The National Institute of Health Stroke Scale (NIHSS) was 1.

Table

Test	Lab result	Reference range
Partial Thromboplastin Time (PTT)	76.4 seconds (H)	(26-36)
LDL	112 mg/dl	(<130 mg/dl)
Antinuclear antibodies (ANA)	ANA titer 1 1:60 Positive	(<1:40 titer Negative)
Anticardiolipin Antibodies (ACLA)	Ig G 62.1 GPL unit/ml	(0.0-15.0; positive > 20)
ACLA Ig M	13.6 MPL unit/ml	(0.0-12.5; positive >20)
Double stranded DNA antibodies (dsDNA abs)	155 IU/ml Positive	(> = 10, positive)
Beta-2 Glycoprotein antibodies Ig G (B2 Ig G)	55.73 SGU Positive	(0.0-20.0)
B2 Ig M	5.15 SMU	(0.0-20.0)
Erythrocyte Sedimentation Rate (ESR)	32 mm/hr (H)	(0-15)
Activated Protein C Resistance (APCR)	4.7 ratio Positive	(ratio > or = 2.1, negative)

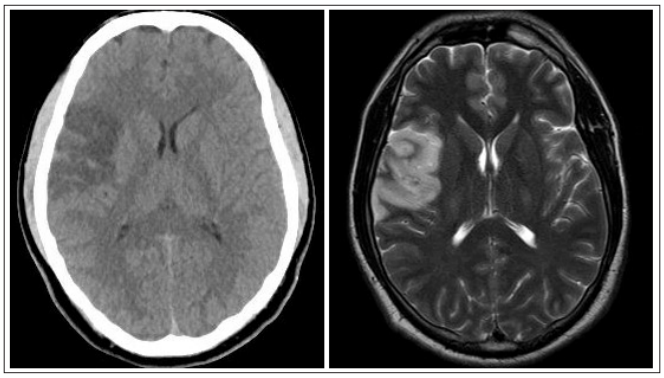

Figure 1: Computed tomography (CT) scan of the brain without intravenous contrast revealed acute to subacute infarct involving the right middle cerebral artery territory without evidence of acute cerebral hemorrhage.

Figure 2: Magnetic Resonance Imaging (MRI) of the brain without intravenous contrast revealed acute right posterior inferior frontal lobe and insula infarct measuring up to 5.6 cm in diameter, mild local mass effect without herniation.

On further imaging, CT angiography of the head and neck revealed a relatively stable infarction involving the right middle cerebral artery territory. CT angiography of the neck was ordered to rule out a possible carotid dissection. Transesophageal Cardiac Echography (TEE) revealed no clots in the left ventricle, non-significant valvular disease, no evidence of shunts and no patent foramen ovale. No rhythm abnormalities were identified on telemetry in the first 24 hours. Based on the patient's imaging and laboratory results he was started on Atorvastatin 80 mg, Aspirin 81 mg daily and lisinopril 5 mg for hypertension.

After three days in the hospital, he was asymptomatic and was discharged with instructions to follow-up as an outpatient (rheumatology/hematology) for a repeat aPL test in 12 weeks.

Discussion

Antiphospholipid syndrome is a rare cause of ischemic stroke and should be suspected in young individuals without cardiovascular-dependent risk factors. The actual frequency of APS in the general population is unknown. One to 5% of healthy individuals have aPL antibodies. It is estimated that the incidence of APS is approximately 5 cases per 100,000 persons per year, and the prevalence is approximately 40-50 cases per 100,000 persons [4]. APS can occur as a primary condition or in the setting of other systemic autoimmune diseases such as SLE [1]. SLE is present in about 35% of patients with APS, with frequent evolution of APS into SLE [2]. Thromboses are a hallmark of APS [2]. The risk of thrombosis increases with the presence of Lupus anticoagulant (LA) [5]. Unfortunately, due to technical issues, we were not able to test the presence of LA in our patient. In addition to antiphospholipid antibodies (aPL), other potential laboratory findings include thrombocytopenia, hemolytic anemia, prolonged activated partial thromboplastin time (aPTT), a history of a false-positive serologic test for syphilis, and low complement levels [2].

Generally, the diagnosis of APS is made in the presence of aPL in the setting of a vascular thrombosis or pregnancy morbidity [1]. The revised Sapporo criteria requires that a patient satisfy at least one clinical criterion related to either a vascular thrombosis or an adverse pregnancy outcome, as well as the presence of one or more specified aPL on two or more occasions at least 12 weeks apart [1]. Our patient was scheduled with hematology-oncology to have a repeat test for aPL in 12 weeks. The mainstay of treatment of acute thromboembolism with APS is anticoagulation, preferably heparin bridged with warfarin rather than Direct oral anticoagulants [3]. For patients with arterial thrombosis, aspirin can be added in selected patients [3]. Dual antiplatelet can be stopped and anticoagulation started if indicated [4]. Our patient had a TIA 2 days prior to arrival and was discharged home without any antiplatelet therapy; if properly assessed at the time of the TIA using the ABCD2 score, He could have received single or dual antiplatelet therapy and reduce the risk of him subsequently having a stroke. TIAs are episodes of stroke symptoms that last only briefly; the standard definition of duration is <24 h, but most TIAs last <1 h [5]. Once the diagnosis of APS was confirmed with our patient, he was started on warfarin and maintained on aspirin. Because prolonged aPTT can be associated with APS such as in our patient, it cannot be used to evaluate therapeutic range prior to bridging to warfarin. Instead, anti-factor Xa assay should be used to monitor heparin anticoagulation prior to bridging [6].

Conclusion

Antiphospholipid syndrome is a rare condition that can present as thrombosis in patients without underlying cardiovascular comorbidities. It is important to keep in mind that APS can cause thrombosis in both the arterial and venous systems. While APS is more often associated with early miscarriage and pregnancy morbidity, it should be ruled out in cases of unexpected thrombotic events in people under the age of 50.

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