Resolving the neurologic status with improvement of the overall skin findings was achieved following head magnetic resonance imaging (MRI) showed multifocal areas of T2 hyperintensities, possibly from lupus vasculitis. In another patient, a Caucasian American female with a history of SLE associated with increased risk of stroke and premature death due to cerebral infarction of arterioles and arteries with fibrin thrombi, occlusion of veins or veins; minimal inflammation; typically lacks leukocytoclastic vasculitis, and causes direct injury and affect the blood-brain barrier, allowing antibodies to enter the nervous system, characterized by a small to moderate perivascular accumulation of mononuclear cells in blood vessels, resulting in small infarcts due to luminal occlusion. Laboratory findings: Positive antiphospholipid antibodies: anti-cardiolipin antibodies (most sensitive), lupus anticoagulant, anti-β2-glycoprotein I antibody (most specific), increased the risk of stroke and seizures, and lupus anticoagulant. Treatment of APS is typically a combination of anticoagulation (e.g. warfarin), antplatelet agents (e.g. aspirin), and antimalarial agents (e.g. hydroxychloroquine). To date, long-term anticoagulation has been the only treatment shown to reduce vascular complications. However, that regimen does not prevent organ deterioration and death in high-risk patients. Treatment underlining cause such as infection and discontinue any responsible drugs. This case highlights the predisposition to thrombosis and vasculitis noted in patients with APS. Clinicians must have a high degree of suspicion for the presence of antiphospholipid antibodies in women with a history of SLE presenting with neurologic symptoms. The presence of antiphospholipid antibodies in the appropriate clinical setting is key to establishing diagnosis. Prolonged anticoagulant therapy is the mainstay of treatment. Low-molecular weight heparin and low-dose aspirin preferred for pregnant patients, and pravastatin may improve pregnancy outcomes when taken at onset of preeclampsia and intrauterine growth restriction. Treatment often includes antplatelet agents and antimalarial agents as well. 

REFERENCES


INTRODUCTION

- Antiphospholipid syndrome (APS) is a rare acquired autoimmune disease, characterized by the formation of autoantibodies to cardiolipin, lupus anticoagulant, and beta-2-glycoprotein I.1
- Affects primarily young women.1
- Predisposes to thrombosis and obstetric morbidity.1
- Most commonly associated with an underlying autoimmune disease such as systemic lupus erythematosus (SLE). Less frequently, occurs in the setting of infection or lymphoproliferative malignancy. In addition, several medications are associated with APS.1
- Empiric anticoagulation, anti-platelets, and antimalarial agents are the treatment of choice in those with concurrent lupus.1
- Featured as a case of antiphospholipid syndrome presenting as discoid lupus erythematosus (DLE), persistent, and cerebral infarctions in a young woman with SLE.

PRESENTATION

- A 26-year-old African American female complaining of a two-week history of progressive, throbbing headache and rash involving her nose and distal extremities following a flu-like illness. Associated myalgia, arthralgia, lightheadedness, blurred vision, and right lower extremity weakness reported.
- Recent history of upper respiratory illness with rhinorrhea, nasal congestion, and shortness of breath relieved by over-the-counter decongestants.
- Denied recent sun exposure, chemical exposures, travel, vaccinations, sick contacts.
- Past medical history included SLE, DLE, arthritis, and chronic lower back pain.
- Home medications included hydroxychloroquine and mycophenolate mofetil.
- Family history, allergies, and social history noncontributory.
- Physical examination revealed scar-like, atrophic, erythematous plaques covered with adherent scales of the nose and malar cheeks (Figure 1). Multiple purpura, tender erythematous to violaceous macules and edematous papules were noted on the Palmer hands and plantar feet (Figure 2).
- Laboratory tests revealed leukocytosis and elevated ESR.
- Renal function tests, coagulation profile, cultures, urine analysis, and chest radiograph were unremarkable.
- Lupus anticoagulant and anti-cardiolipin antibodies positive.
- Lumbar puncture and cerebral spinal fluid (CSF) analysis negative.
- Head computed tomography (CT) showed nonspecific leukoencephalopathy (Figure 3).
- Head magnetic resonance imaging (MRI) showed multifocal areas of T2 hyperintensities throughout the subcortical and deep white matter, multiple areas of restricted diffusion bilaterally suggestive of multiple infarctions, possibly from lupus vasculitis (Figure 4).
- Findings were consistent with APS.
- Resolution of her neurologic status with improvement of her overall skin findings was achieved following initiation of high-dose prednisone, aspirin, and anti-coagulation.

DISCUSSION

- The antiphospholipid syndrome is an autoimmune disease characterized by the presence of circulating antiphospholipid antibodies that result in vascular thrombosis and obstetrical complications.2
- Occurs mainly young women1
- Predisposition to thrombosis, the major complication of APS5
- Clinical presentation
- History of vascular thrombosis, premature birth, recurrent miscarriage, and labile blood pressure4
- Livedo reticularis is the most common cutaneous finding1
- Other cutaneous findings include atrophic blanche, leg ulcers, vasculitis, pseudovasculitis, digital gangrene, cutaneous necrosis, splinter hemorrhages, cyanosis, perniosis, and retiform purpura suggestive of occlusion.
- Systemic features include DVT, PE, stroke, renal infarct, myocardial infarction, arthritis, and seizures2
- Often with autoimmune disorders, such as SLE (most common), rheumatoid arthritis, and ulcerative colitis.1
- SLE associated with increased risk of stroke and premature death due to cerebral infarction5
- Stroke has been reported in up to 19% of patients with SLE4
- Precipitants may include surgery, infection (e.g. HIV, hepatitis C), and medications (e.g. hydrocholorothiazide, oral contraceptives, ace inhibitors)3
- Pathology
- Occlusion of arteries and arterioles with fibrin thrombi, occlusion of veins or veins; minimal inflammation; typically lacks leukocytoclastic vasculitis5
- Vasculopathy may cause direct injury and affect the blood-brain barrier, allowing antibodies to enter the nervous system, characterized by a small to moderate perivascular accumulation of mononuclear cells in blood vessels, resulting in small infarcts due to luminal occlusion5
- Laboratory findings
- Positive antiphospholipid antibodies: anti-cardiolipin antibodies (most sensitive), lupus anticoagulant, anti-β2-glycoprotein I antibody (most specific)2
- Increase the risk of stroke and seizures2
- Associated with an increased prevalence of abnormal findings on MRI4
- False-positive syphilis serology2
- Sapporo criteria for diagnosis of APS were revised in 2006 - one clinical finding along with one positive laboratory criterion must be present4
- Treatment of APS is typically a combination of anticoagulation (e.g. warfarin), antiplatelet agents (e.g. aspirin), and antimalarial agents (e.g. hydroxychloroquine).1
- To date, long-term anticoagulation has been the only treatment shown to reduce vascular complications. However, that regimen does not prevent organ deterioration and death in high-risk patients.2
- Treat underlying cause such as infection and discontinue any responsible drugs.8

CONCLUSION

- This case highlights the predisposition to thrombosis and vasculitis noted in patients with APS.
- Clinicians must have a high degree of suspicion for the presence of antiphospholipid antibodies in women with a history of SLE presenting with neurologic symptoms.
- The presence of antiphospholipid antibodies in the appropriate clinical setting is key to establishing diagnosis.
- Prolonged anticoagulant therapy is the mainstay of treatment.
- Low-molecular weight heparin and low-dose aspirin preferred for pregnant patients, and pravastatin may improve pregnancy outcomes when taken at onset of preeclampsia and intrauterine growth restriction.8
- Treatment often includes antiplatelet agents and antimalarial agents as well.9

REFERENCES