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Recurrent Cerebrovascular Accident in Antiphospholipid Syndrome - A Case Report

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ABSTRACT

Antiphospholipid syndrome is an autoimmune disorder with arterial events, venous thromboembolism, and obstetric morbidity associated with antiphospholipid antibodies. Cerebral arteries are the most common site for arterial thrombosis causing stroke (13.1%). The occurrence of arterial thrombosis carries a poor predictive value, given the high risk of recurrence. The treatment with apixaban is controversial due to limited studies; thus, Vitamin K antagonists are recommended for thrombotic antiphospholipid syndrome with an INR between 2.0 and 3.0.

Keywords: Apixaban; Antiphospholipid; Cerebrovascular accident; Thrombosis; Warfarin; Stroke

INTRODUCTION

Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by recurrent pregnancy complications and/or thrombosis (arterial, venous, microvascular) due to antiphospholipid antibodies (aPL) such as lupus anticoagulant, anti-β2-glycoprotein 1 and anticardiolipin.[1] The overall annual incidence of APS is about 2 persons per 100,000 per year, and the estimated prevalence is 50 per 100,000.[2] In a recent population-based study, the mean of APS diagnosis reported is around 50 years, while the study done in the US showed the peaked incidence in males is 55-64 and more than 75 years in women.[3] 10% of arterial events, venous thromboembolism, and obstetric morbidity are associated with Antiphospholipid antibodies (aPL). The most common manifestations of APL are deep vein thrombosis (31.7%), thrombocytopenia (21.9%), livedo reticularis (20.4%), stroke (13.1%), pulmonary embolism (9.0%), fetal loss (8.3%), transient ischemic attack (7.0%), and hemolytic anemia (6.6%).[4]

Cerebral arteries and deep veins of the lower extremities are the most frequent sites of arterial and venous thrombosis in APS, respectively. Vitamin K antagonists are recommended for thrombotic episode antiphospholipid syndrome with an INR between 2.0 and 3.0 or 3.0 and 4.0.[5,9]

CASE SUMMARY

A 66-year-old male with a past medical history of Cerebro-vascular accident (CVA) and pulmonary embolism secondary to the antiphospholipid syndrome, peripheral vascular disease hypertension, dyslipidemia, chronic obstructive pulmonary disease (COPD) presented with sudden onset of left-sided weakness and dysarthria associated with right arm tingling sensation and right-sided drooping of the face. His NIH score on presentation was 6. He was compliant with his medications, including apixaban (5mg BD), clopidogrel, atorvastatin, and albuterol inhaler. The patient smokes one pack of cigarettes daily and does not use smokeless tobacco, alcohol, or illicit drugs.

He had 24.15kg/m^2 with a blood pressure of 159/78. On examination, the patient was aphasic but was able to understand and had drooping of the right nasolabial fold. Neurological examination revealed diminished crude touch and power (4/5) on both upper and lower right limbs and normal power on the left side. His cardiovascular, respiratory, musculoskeletal, and abdominal findings were unremarkable.

On laboratory investigation, he had hemoglobin (13.2 gm/dl), platelet (124), PTT on heparin (27.4 seconds), INR (1.02), blood urea nitrogen (32mg/dl), creatinine (1.9 mg/dl), cholesterol (151 mg/dl), HemoglobinA1C (5.9 %). In 2019, laboratory investigation was positive for the lupus anticoagulant (1.59) and Cardiolipin Ab IgG (17). His current CT scan of the head, echocardiography, and CT neck angiography were unremarkable. However, an MRI of the brain revealed small areas of acute infarcts within the superior division M2 segment of the left Middle Cerebral artery with no acute intracranial hemorrhage or hemorrhagic transformation (Figure1). Hence, consultation with the neuro medicine was done, who recommended switching apixaban to warfarin with a goal INR 2-3 as per the 16th International Congress on Antiphospholipid Antibodies Task Force Report on Antiphospholipid Syndrome Treatment Trends.[6] The patient's condition is currently well maintained with the introduction of warfarin.

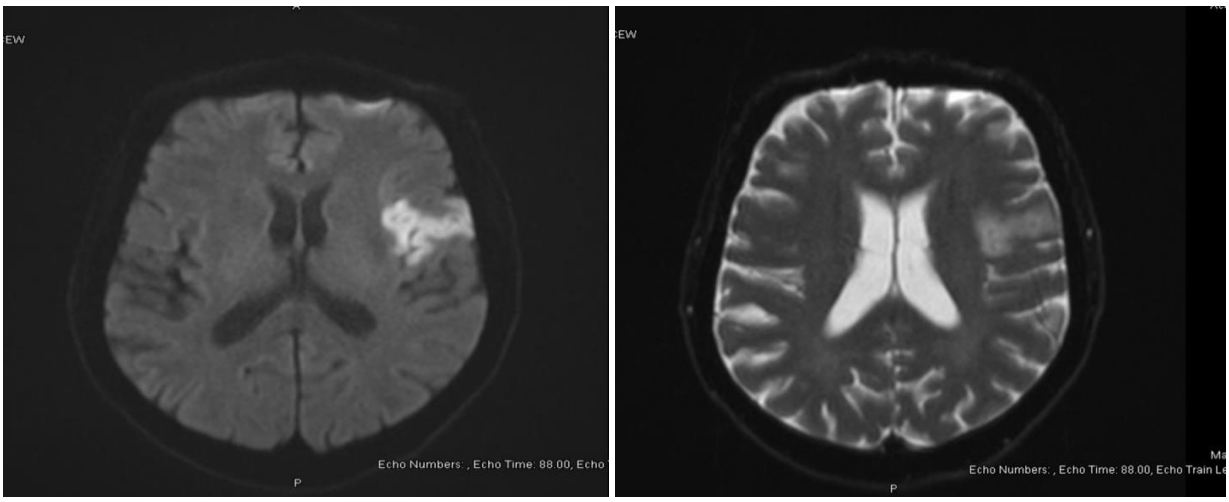

Figure1: MRI brain showing small areas of acute infarcts within superior division M2 segment of left Middle Cerebral artery.

DISCUSSION

In the presence of antiphospholipid antibodies (aPL), APS is an autoimmune acquired thrombophilia characterized by recurrent arterial and venous thrombosis and pregnancy morbidity. These aPLs are pathogenic and play a crucial role in thrombosis. The thrombus formation in APS patients is proposed as a "two-hit" thrombosis model. A "first hit" injury to the endothelium is required for a "second hit" that promotes thrombus development.[7] Furthermore, smoking might cause endothelial damage and enhance pro-thrombotic susceptibility in lupus anticoagulant patients. Our patient had two antibodies, lupus anticoagulant and anticardiolipin, and smoked one pack per day, which may have raised the risk of thrombus formation, resulting in a second episode of cerebrovascular accident (ischemic stroke) despite taking apixaban and clopidogrel. A systematic study found that a positive lupus anticoagulant (odds ratio >10) increases the risk of thromboembolism more than cardiolipin antibodies.

APLS can result in arterial and/or venous thrombosis in any organ system. APLS-related thrombotic events can develop in the absence of a prior risk of thrombosis. Arterial thrombosis can affect any size artery (from the aorta to small capillaries). Transient ischemic episodes (TIAs) or ischemic stroke are the most prevalent arterial manifestations of APLS. Because of the increased probability of recurrence in these patients, the presence of arterial thrombosis has a poor predictive value.[7]

Vitamin K antagonists are recommended for thrombotic episode antiphospholipid syndrome with an INR between 2.0 and 4.0. Because patients with APS are typically prescribed warfarin for life, the problem arises when people prefer direct oral anticoagulant (DOAC) or experience VTE recurrence while on warfarin. Apixaban may thus be considered in individuals who refuse warfarin medication, are warfarin intolerant, or have poor anticoagulant control despite adherence.[8] When compared to warfarin, DOACs have the advantages of being prescribed at set doses, having predictable anticoagulant effects, not requiring routine anticoagulation monitoring, having fewer drug interactions, and not causing any gastrointestinal problems. These qualities are intriguing for thrombotic APS

patients who typically require lifelong anticoagulation. A case series by Al Sulaiman K et al. reported some cases with apixaban 5 mg twice daily with no recurrent thrombotic and bleeding events.

Thus, in patients with high levels of warfarin aversion (e.g., labile INR, numerous recurrent thrombotic events), a full therapy discussion addressing advantages and thrombotic recurrence risk with apixaban remains an important consideration for apixaban introduction. However, few studies reported the role of apixaban in APL as fatalistic. Due to limited available data, the present role of apixaban in treating APS remains unknown, and further studies are required.

The 16th International Congress on Antiphospholipid Antibodies Task Force report on antiphospholipid syndrome treatment trends suggested DOAC should be avoided in APS patients with arterial thrombosis, small vessel thrombosis, and recurrent thrombosis and recommended Vitamin K antagonist as the first line anticoagulation.[6] Patients with moderate-to-high-risk aPL profiles are often managed with warfarin (target INR 2-3), with or without low-dose aspirin.

CONCLUSION

APL presents with various systemic manifestations affecting multiple organs. However, the recommended treatment for arterial thrombosis due to APL is the vitamin K antagonist, warfarin.

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