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Case Report

Subacute Stroke and Myocarditis Associated with Antiphospholipid Syndrome

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Abstract

Ischemic stroke is one of the most common complications of the antiphospholipid syndrome (APS). The mechanisms of ischemic stroke could be atherothrombotic and/or embolic. Transesophageal echocardiograms are strongly recommended in APS patients with ischemic stroke because of the high yield of valvular abnormalities. For management, it's recommended warfarin or another vitamin K antagonist for arterial thrombosis out-side the cerebral vasculature. For older patients with strokes and a low risk profile (low title of aCl antibodies), aspirin alone may be as effective as warfarin; in patients with moderate to high risk antibody profiles, warfarin is indicated (target INR, 2 to 3), with or without low-dose aspirin.

Keywords: *Antiphospholipid Syndrome (APS), Anti Beta2 Glycoprotein I, Systemic Lupus Erythematosus, Lupus Anticoagulant, Anticardiolipin Antibodies*

Introduction

APS is the most common cause of acquired thrombophilias, which cause arterial and venous thrombosis. Its clinical presentation is heterogeneous. Laboratory criteria are defined by the detection of some antibodies: lupus anticoagulant, anticardiolipin and/or anti beta2 glycoprotein I, in two determinations separated by 12 weeks. The risk factor depends on the association of these three determinations (positivity in 1, 2 or 3 antibodies), and its titers. Ischemic stroke is one of the most frequent arterial complications of APS. Data suggests that more than 20% of strokes in those under 45 years of age are associated with APS. Next in prevalence is cardiac involvement, with thickening of the heart valves with a prevalence of 30%. For these cases, the prevalence of myocardial alterations suggestive of microvascular thrombosis and myocarditis must be evaluated, and their early detection by cardiac resonance in APS. (1-5)

Case Report

A 57-year male, with a history of high blood pressure, Lupus with kidney involvement, deep vein thrombosis (anticoagulated for 6 months), was admitted in the hospital due to episodes of anomia and disorientation lasting 3 weeks. On physical examination, there were no sensory motors or cranial nerve deficits. Preserved taxia. A brain MRI was performed showing left thalamic ischemic lesion in a subacute stage (Fig 1).

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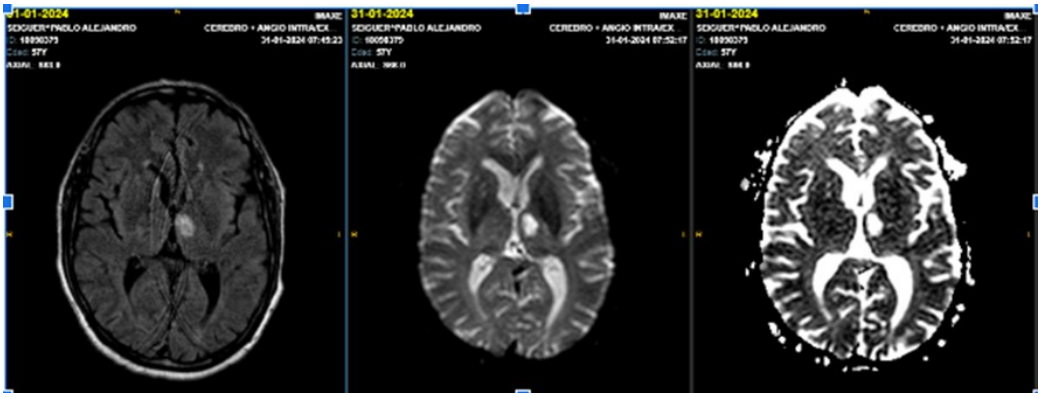

Fig 1: *Brain MRI (Flair, DWI, ADC) subacute ischemic lesion is evident in the left thalamus*.

A laboratory test was performed, with a positive lupus inhibitor and positive high titers Igm anticardiolipin antibodies. A transthoracic echocardiogram was performed, which reported anteroseptal and apical hypokinesis, with ejection fraction 34%, and a transesophageal echocardiogram that ruled out the presence of intracardiac thrombus. Coronary angiography did not show endoluminal lesions. A cardiac MRI with gadolinium was requested, which revealed anteroseptal subepicardial uptake, interpreted in conjunction with cardiology as probable myocarditis with hypokinesis in the left ventricle secondary to APS. Treatment with heparin started. At 12 weeks, a repeat laboratory confirmed the presence of positive antibodies for SAF.

Discussion

Antiphospholipid syndrome is a systemic autoimmune disease defined by arterial and/or venus thrombotic events that occur in patients with persistent antiphospholipid antibodies. Nonthrombotic manifestations include valvular heart disease, livedo, nephropathy, thrombocytopenia, hemolytic anemia, and cognitive dysfunction. Antiphospholipid syndrome is often associated with systemic lupus erythematosus (SLE); however, it commonly occurs without other autoimmune manifestations (primary antiphospholipid syndrome).

The positivity of the antibodies without clinical events, does not make a diagnosis. It's well known that 10% of healthy people are positive for anticardiolipin antibodies, and 1% positive for lupus anticoagulant approximately. In patients who have SLE, with persistent moderate-to-high-risk antiphospholipid-antibody profiles, 25% are associated with an increased risk of clinical thrombotic events.

Stroke is the most common arterial event in patients with APS. Cardiac involvement is characterized by valve vegetations or thickening. It can be diagnose with transesophageal echocardiography when there is a valve thickness >3 mm, thickening of the proximal or middle portion of the leaflet, or irregular nodules on the atrial face of the edge of the mitral valve, the vascular face of the aortic valve, or both.

As we mentioned before, a unique positive aPL test is not clinically significant per se. LA testing is a three-step functional coagulation assay to detect aPL. The LA test correlates better with clinical events than do aCL and anti-β2GPI tests. The determination of LA is performed in a three-step coagulation test and correlates better with clinical events than with the determinations of the other antibodies. There may be false positives for LA, especially in patients treated with warfarin or other anticoagulants. Profiles have been defined based on the titer and association between these three antibodies (aCL and anti-β2GPI (IgG, IgM or IgA) and LA) to stratify the risk of clinical events in these patients. A high risk profile (LA positive with or without a moderate-high titer of aCL or anti-β2GPI), moderate risk (LA negative with a moderate to high titer of aCL or anti-β2GPI) and low risk (LA negative with a titer low aCL or anti-β2GPI).

For management, it's recommended warfarin or another vitamin K antagonist for arterial thrombosis out-side the cerebral vasculature. For older patients with stroke and a low risk profile (low title of aCl antibodies), aspirin alone may be as effective as warfarin;in patients with moderate to high risk antibody profiles, warfarin is indicated (target INR, 2 to 3), with or without low-dose aspirin. The risk of new neurovascular events in these patients it's not well established.

For older patients with stroke and a single test showing a low titer of anticardiolipin antibodies, aspirin alone may be as effective as warfarin; however, patients with moderate-to-high-risk antiphospholipid-anti- body profiles are often treated with warfarin (target INR, 2 to 3), with or without low-dose aspirin. In patients with recurrent thrombosis despite therapeutic dose anticoagulant therapy, adding hydroxychloroquine can be considered. (1-5)

Conclusion

In patients with APS, the stroke mechanism is usually atheroembolism or cardioembolic and may be associated with intracardiac and valvular thrombus anomalies, which are predictors of poor outcome. A higher frequency of patent foramen ovale and atrial septal aneurysm has been described in patients with stroke and APS, which is why they should be ruled out.

Those who present a parallel cardiac involvement usually have a high rate of embolic events, close to 40%. Regarding treatment, the guidelines published on the treatment of stroke by the American associations in 2019 (AHA) recommend that for patients with ischemic stroke or TIA, who meet the criteria for APS, anticoagulant treatment can be considered according to the risk of episodes, recurrent thrombosis and hemorrhage (class IIb; level of evidence C).

Conflicts of Interest

The authors declare no conflicts of interest.

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