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Case Report

Primary Antiphospholipid Syndrome: A Case Report of Embolic Ischemic Stroke and Cortical SAH due to Libman-Sacks Endocarditis

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Abstract

This is report of 27 years old lady admitted with acute onset aphasia, right sided hemiparesis and facial paresis without of previous history of Cardiac or other atherosclerotic risk factors, abortions, rheumatologic disorder, thrombotic events. Ischemic in right MCA territory and right frontal lobe signal change in favor of cortical subarachnoid hemorrhage were the findings in MRI.

Positive Antiphospholipid antibody associated with mitral valve vegetation, suggestive of Nonbacterial thrombotic endocarditis (NBTE) in Trans-Esophageal Echocardiogram (TEE) were the positive finding in evaluation. So with diagnosis of NBE in the setting of antiphosholipid antibody, anticoagulant therapy was started. Screening for Malignancy were negative.

NBTE also known as Libman-Sacks Endocarditis, Marantic Endocarditis, is a rare condition according to autopsy series ranging from 0.9 to 1.6 percent. It has been reported in every age group, most commonly affecting patients between the fourth and eighth decades of life with no sex predilection. Mucus secreting malignancy, systemic lupus erythematosus and APS syndrome are the common etiologies.

Keywords: Juvenile Stroke; Antiphospholipid Syndrome; Libman-Sacks Endocarditis; NBTE

Case Presentation

This is report of 27 years old lady admitted with acute onset aphasia, right sided hemiparesis and facial paresis without of any previous history of Cardiac or other atherosclerotic risk factors, abortions, rheumatologic disorder, thrombotic events since four days ago. Gestational hypertension, seizure attacks since childhood, with last episode 6 months ago were the important past history. She did not experience trauma or falling prior to her symptoms.

Neurological examination revealed disorientation with global aphasia, facial palsy and right hemiparesis with NIHSS score of 14.

Hemodynamic parameters, EKG, Transthoracic Echocardiography were normal.

MRI in DWI sequences showed restriction in Lt MCA distribution at temporoparietal and thalamus areas with linear hyper signal in FLAIR sequences suggestive of cortical SAH in Right frontal area.

Brain MRV and CT-Angiogram of cervico-cephalic vessels were normal.

Evaluation for of cryptogenic stroke showed high ESR, CRP, positive Antiphospholipid Ab, Anticardiolipin Ab IgG and Anti B2-Glycopeotein.

Transesophagial echogardiogram revealed mobile mass about 1.6*1.3 cm attached to atrial side of PMVL, suggestive of NBTE. Rheumatologic consultation approved the diagnosis of primary antiphospholipid syndrome with nodocumentry criteria for SLE. Anticoagulant with Heparin that followed with warfarin with INR target of 2.5-3.5 was the main treatment.

Confuional state, partial Broca's aphasia, normal power with NIHSS of 6, reduced size of the mobile mass in TEE were the major finding in three weeks follow up.

Discussion

The presence of mobile mass in TEEC in association of positive antiphospholipid antibody was the hallmark of libman-sacks endocarditis as a predisposing factor for arterial embolic stroke in a young lady.

NBTE is a rare condition that refers to a spectrum of noninfectious lesions of the heart valves, in the setting systemic of lupus erythematosus [1], advanced malignancy (1.25 versus 0.2 percent in normal population) in autopsy, mostly in those with adenocarcinoma (e.g. lung, colon, ovary, biliary and prostate) (2.7 versus 0.47 percent in other cancers) and highest rates observed in patients with mucin-secreting and pancreatic adenocarcinoma (10.34 percent versus 1.55 in other adenocarcinomas) [2].

In patients with systemic lupus erythematosus, observational studies using transthoracic echocardiography have reported prevalence rates of 6 to 11 percent, with higher rates (43 percent) when transesophageal echocardiography was performed [3].

Kitagawa et al showed Association of cerebrovascular events and Antiphospholipid (APL) antibodies in 5.6% of lupus patients. LAC was detected in 38% and ACL in 43% of studied patients with stroke [4]. Also contribution of libman-sacks endocarditis to ischemic stroke have been reported in two cases [5].

Previous studies demonstrated association of SAH with Lupus, but connection of antiphospholipid antibodies in lupus or presence of APS was not evident in studied patients [4].

Presence of Non-traumatic cortical SAH in the absence of arteriovenous malformations, cortical venous or dural sinus thrombosis, and distal and proximal arteriopathies (RCVS, vasculitis, mycotic aneurysms, Moyamoya, or severe atherosclerotic carotid disease) was the important manifestation in this patient [6].

Antiphospholipid syndrome is reported as an etiology of cryptogenic stroke in 20-25 percent of patients under the age of 50 [7], but association of valvular vegetations occurring is rare in 6–10%. [8]. Valvular surgery should be considered in patients with large mobile vegetations, significant valvular dysfunction or recurrent embolic events [3].

As warfarin therapy seems to be a promising therapy for resolution of vegetation in APS patients [8]. Initiation of oral anticoagulation is mandatory in APS, after the first event. In the case of an initial venous event, a low dose (target INR between 2.0 and 2.6) may suffice, but intermediate dose anticoagulation (target INR between 2.6 and 3.0) is recommended after an initial arterial or recurrent venous events [4]. As Moderated to high intensity anticoagulation is also necessary in patients with cardiac embolism, so this lead us to monitor INR with target of 3-3.5 [8]. Therapy must be prolonged, usually lifelong, but there are no definite guidelines for the exact duration. Aspirin use is becoming increasingly fre-

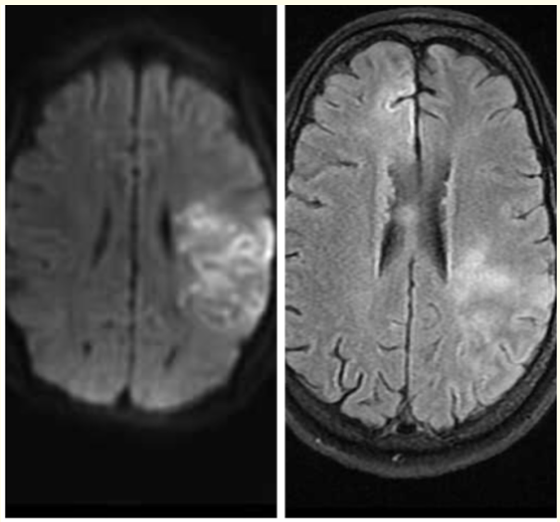

Figure 1: A: DWI sequence in favor of acute ischemic stroke, B: FLAIR sequence of linear cortical hypersignal area in favor of cortical SAH

quent in the secondary prevention of embolic phenomena in these patients, but more data is in favor of beneficial effect of this drug in preventing the arterial events rather than venous events [9]. Corticosteroid and other immune-modulating drugs may be beneficial in preventing development of cardiac lesions in APS secondary to SLE [10], but not in primary APS [4].

Conclusion

In patients with juvenile stroke, especially females, primary APS should be considered as an important risk factor, even without accompanying other connective tissue disorders, such as SLE. This significance is because in addition to its direct effect of APS on evolving ischemia presence of emboli in the setting of libman-sacks endocarditis leads us to anticoagulation with INR target of 3-3.5 in the setting of primary APL without use of immunosuppressive drugs.

Competing Interests

All authors declare no conflict of interest. This research did not receive any specific grant from any funding agencies in the public, commercial or not-for-profit sectors.

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