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Case Report

The Difficult Diagnosis: Late Onset Antiphospholipid Antibody Syndrome

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Abstract

Antiphospholipid antibody syndrome is defined by the presence of recurrent thrombosis with and without recurrent miscarriages due to the presence of certain antiphospholipid antibodies including lupus anticoagulant, anti-cardiolipin and anti-β2GP1 antibodies. It is usually seen in the younger patients, and is rarely described in the elderly population. In younger patients with stroke, APS is considered as a possible etiology, however in elderly, APS has low probability due to multiple pre-disposing risk factors of stroke. We reported a case of 67-year-old female, with history of multiple strokes in the past, presented with slurred speech, facial droop and left arm weakness. CT head reported cortical hyper-intensity involving anterolateral frontal lobe consistent with acute ischemic R sided CVA. Her recent laboratory results were obtained from her clinic records and she was found to have elevated lupus anticoagulant and elevated levels of anti-cardiolipin antibodies. The hypercoagulable cause of her recurrent cerebrovascular accident was found and patient was diagnosed with late onset antiphospholipid antibody syndrome. She was discharged on warfarin anticoagulation along with other home medications.

Background

Antiphospholipid antibody syndrome is defined by the presence of antiphospholipid antibodies (ApL) such as lupus anticoagulant (LAC) [1], anti-cardiolipin (aCL) and anti-β2GP1 antibodies against beta-2-glycoprotein resulting in recurrent arterial and venous thrombotic events. It may also result in recurrent miscarriages. Among several types of anti-phospholipid antibodies [2], LAC has been shown to be the strongest risk factor for thrombosis. The syndrome is usually seen before the fifth decade of life. It has been rarely described in the elderly population. In younger patients with stroke, APS is considered as an etiological risk factor, however in patients above 70 years of age, APS has low probability due to multiple pre-disposing risk factors of stroke in this patient population group, making the diagnosis of stroke from APS unlikely.

Case Presentation

A 67-year-old female admitted with the complain of slurred speech, facial droop and left arm weakness with numbness. She provided the past medical history significant for two ischemic strokes, first one in 2004 and the second ischemic stroke three months ago, resulting in residual left arm weakness. Rest of the past medical history included essential hypertension and hyperlipidemia. Current medications include amlodipine, aspirin, clopidogrel and simvastatin. Patient denied confusion, blurred vision, dysphagia, and headache.

On physical examination patient was found alert and well oriented, with left arm weakness with power 4/5 in left arm, dysarthria and left UMN facial palsy with mild-to-moderate sensory loss (NIH scale of 4). Patient however had normal comprehension along with normal motor skills and cranial nerves. Initial lab work was normal except for low Hb, elevated pTT and elevated fibrinogen levels. CT head reported cortical hyper intensity involving anterolateral frontal lobe consistent with acute ischemic R sided CVA (Figure 1). MRI brain showed diffusion weighted abnormality in the right frontal lobe due to acute ischemic changes with no evidence of hemorrhage. MRA neck study for facial asymmetry and arm weakness was negative for any arterial abnormality. Echocardiogram and carotid Doppler were also unremarkable.

Patient was started on tPA therapy and showed improvement in dysarthria and left arm numbness. During her hospitalization stay, patient mentioned remote history of recurrent first trimester abortions in her 20's and several premature deaths in her maternal family. Her recent laboratory results were obtained from her clinic records and she was found to have elevated lupus anticoagulant and elevated levels of anti-cardiolipin antibodies. The hypercoagulable cause of her recurrent cerebrovascular accident was found and patient was diagnosed with late onset antiphospholipid antibody syndrome [3]. She was discharged on warfarin anticoagulation along with other home medications. The patient later underwent physical rehabilitation and reached her baseline functionality of mild left hand weakness.

Discussion

Antiphospholipid antibody syndrome is mainly seen in the younger patient population. There are few studies in literature documenting APS in elderly. A case reported by Delgado et al, mentioned a case of a 77-year-old female with past medical history of atrial fibrillation, on warfarin, admitted with language difficulties secondary to left MCA stroke. Patient had a history of multiple ischemic stroke episodes in the past 3 years and the occurrence of these strokes was attributed to the Cardioembolic etiology. On her fourth stroke, she was evaluated for APS and complete blood analysis showed presence of anticardiolipin, anti-b2-glycoprotein antibodies, and lupus anticoagulant [4]. Few other studies in the literature support

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Figure 1: CT Head findings reported old infarct and negative for any acute intracranial bleed.

these findings. Euro-Phospholipid project group conducted a cohort of 1000 patients. The study reported 12.7% of patients with APS onset after the age of 50 (referred as older-onset antiphospholipid antibody syndrome) [5,6]. These patients with older onset APS were mostly male, and had the history of more strokes and angina episodes than rest of the participants. The study reported 19.8% cases with stroke, 2.5% with multi-infarct dementia and stroke was called as the third cause of death (13%) among the participants [7]. An interesting study by Cherif et al, describe the late onset of antiphospholipid antibody syndrome in the elderly population group. The retrospective study reported seven out of 62 patients showing different features of thromboembolism. When tested, these seven patients fulfilled the criteria of APS [8]. The study further explains possible impairment in the immune system related to patient's age. APL antibodies have been reported as positive in patients with cancer, liver failure, giant cell arteritis and certain infections. However, the presence of Acl is high in the elderly population. One study was conducted on 64 patients and the results showed 50% positivity of aCL antibodies in patients above the age of 80 [9-11]. A study by Richaud-Patin supported these findings by reporting 63% positivity of Acl and 31.8% of anti-B2GP1 in their elderly population group. However, there are different studies in the literature regarding the occurrence of thrombotic events among elderly with positive acl antibodies. While some studies do not show any support in relating the presence of aCL in vascular events, others regard aCL antibody as an independent risk factor for the thromboembolism in elderly with positive acl antibodies. A study by Tanne et al observed high association of cerebrovascular events and increased mortality among patients with higher than 20-40 GPL levels [12]. Other studies have demonstrated elevated levels of aCL antibodies in patients with multi-infarct dementia and stroke.

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