Pneumonia unmasking APLA syndrome

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Case Report

Abstract: Antiphospholipid antibody syndrome can mimic various other illnesses. We present a patient who presented with features suggestive of consolidation but actually had pulmonary embolism. Despite anticoagulation, she developed cortical vein thrombosis within a short span of time. She improved with continued treatment. Timely clinical suspicion aided quicker recovery and reduced financial burden by avoiding excessive investigations.

Keywords: APLA, pulmonary embolism, cortical vein thrombosis, procoaguable state, antiphospholipid antibody.

1. Introduction

Antiphospholipid antibody (APLA) syndrome has protean manifestations. It can masquerade as various other illnesses and a high index of clinical suspicion is often needed to diagnose APLA. We present a patient who initially was thought to have pneumonia, but diagnosed to have APLA. It was interesting to note how she had two major manifestations of the illness of a span of 20 days.

2. Case Report

A 32-year old woman presented with acute onset left sided chest pain. She had no cough, breathlessness or fever at presentation. The pain subsided with oral analgesics but she developed fever on day 15 of the onset of chest pain. A chest x-ray taken suggested left basal consolidation and she had been treated with a course of antibiotics. She had an ESR of 150mm in the 1st hour, a normal baseline aPTT and INR. However in view of persisting symptoms, a CT scan of the chest was done, and it revealed left pulmonary arterial thromboembolism with air space opacities in the left lower lobe (Fig 1). She was treated with intravenous heparin and discharged on oral dabigatran once symptoms subsided. Three days later she developed severe left sided headache; CT brain revealed left sided transverse sinus thrombosis (Fig 2, 3). She was again treated and was investigated for procoaguable states. Tests for c-ANCA and p-ANCA were negative, and anti-cardiolipin antibody test was also negative. Antinuclear antibody profile was positive for SS-A. Lupus anticoagualant was positive (by dilute Russell viper venom method). She improved with continued anticoagulation and is on regular follow up.

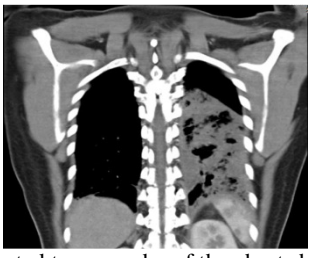

Figure 1: Computed tomography of the chest showing air space opacities in the left lower lobe of the lung due to left pulmonary arterial thromboembolism

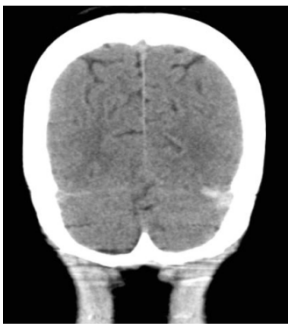

Figure 2: CT brain coronal view showing left transverse sinus thrombosis

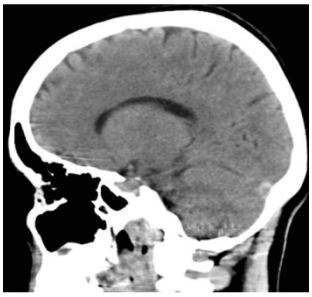

***Figure 3*:** CT brain sagittal view showing left transverse sinus thrombosis

3. Discussion

Antiphospholipid antibodies (APLAs) are abnormal autoimmune proteins in blood that result in a procoaguable state. APLAs include anticardiolipin antibodies, lupus anticoagulant, anti β2 glycoprotein I, antiphosphatidyl serine, antiphosphatidyl ethanolamine, antiphosphatidyl inositol and antiprothrombin; and each of these may be of the IgG, IgA or IgM types. Of these, anticardiolipin antibodies are more commonly found and higher levels increase the risk of clot formation in vivo.

However, a patient with lupus anticoagulant has a greater risk of clot formation than one with anticardiolipin antibodies [1]. It is important to note that while APLAs prolong the time taken for clot formation in vitro (by interfering with phospholipids needed for normal clot formation), they increase tendency to clot in vivo; this may be due to interaction with platelets and due to interference with the anticoagulants proteins C and S. The lupus anticoagulant test involves mixing the individual's blood with phospholipids and measuring the time taken for clot formation. This test may be DRVVT-based (dilute Russell viper venom time) or LA-PTT-based (lupus anticoagulant sensitive partial thromboplastin time). APLA positivity of an individual may imply that he or she had antibodies to one or more types of phospholipids. A person is said to have the APLA syndrome if there is a presence of one clinical event and two positive laboratory tests (6 weeks apart) based on Sapporo criteria [2]; the clinical events include deep vein thrombosis, pulmonary embolism, retinal vein thrombosis, central sinus thrombosis, mesenteric / hepatic / portal vein thrombosis, stroke, myocardial infarction, limb ischemia due to arterial thrombosis, retinal / mesenteric arterial thrombosis, and pregnancy loss or placental insufficiency; the tests include lupus anticoagulant test and high levels of IgG / IgM anticardiolipin antibodies. Our patient had two clinical events and a positive lupus anticoagulant test. When a patient has a co-existing autoimmune condition like systemic lupus erythematosus or rheumatoid arthritis, and is positive for APLA syndrome by Sapporo criteria, he or she is said to have secondary APLA syndrome. APLA syndrome has no known risk of genetic transmission. While most presentations involve one of the above mentioned clinical manifestations, rarely multiple organ systems may be involved within a week [3]. This is then called catastrophic APLA syndrome. Hence, the importance of early diagnosis cannot be overemphasized. Treatment options include anticoagulants, glucocorticoids, plasmapheresis, and immunosuppression. While the risk of a second clinical event is 3% to 10% over three years with anticoagulation, it rises to 10% to 29% per year if anticoagulants are stopped. While warfarin, acenocoumarol or dabigatran are preferred to prevent repeated venous events, the recommended drugs to prevent repeated arterial thrombosis include aspirin, clopidogrel or aspirin combined with dipyridamole. Patients diagnosed with secondary APLA syndrome benefit from immunosuppression using glucocorticoids or drugs like cyclophosphamide, azathioprine, hydroxychloroquine, or rituximab. IVIg may be tried in patients with catastrophic APLA syndrome.

4. Conclusion

The various manifestations of procoaguable states should always be kept in mind when patients have persistent breathlessness or in case of young strokes. Timely clinical suspicion helps us avoid excessive investigations and aids in earlier diagnosis and treatment.

References

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