



Massive Pulmonary Embolism Secondary to Anti-cardiolipin Antibody Syndrome

Dr Jaleel Ahmed Farooq Mohammed

General Practice Consultant – Warwickshire, UK

Date of Submission: 25-09-2020

Date of Acceptance: 5-10-2020

ABSTRACT: Antiphospholipid antibody syndrome, also known as Hughes syndrome, is a hypercoagulable disorder that increases the risk of recurrent vascular thrombosis. We present a case of 45-year-old female who developed massive bilateral pulmonary emboli further work-up of the patient revealed that she had anticardiolipin antibody syndrome

KEYWORDS: Pulmonary embolism, anticardiolipin antibody syndrome, Antiphospholipid syndrome, D-dimer,

I. INTRODUCTION

The antiphospholipid antibody syndrome is a disorder characterised by multiple different antibodies that are associated with both arterial and venous thrombosis.

There are three primary classes of antibodies associated with the antiphospholipid antibody syndrome: (1) anticardiolipin antibodies, (2) the lupus anticoagulant, and (3) antibodies directed against specific molecules including a molecule known as beta-2-glycoprotein.

In 1983 Hughes¹ described the association between the antiphospholipid antibodies and arterial as well as venous thrombosis, and in 1985 proposed this to be a distinct entity, labelling it as anticardiolipin syndrome. However, Harris et al² in 1987 re-named it as antiphospholipid syndrome.

It can occur within the context of several diseases, mainly autoimmune disorders, and is then called secondary antiphospholipid syndrome. However, it may also be present without any recognisable disease, or the so-called primary antiphospholipid syndrome.

Rarely, patients with antiphospholipid syndrome may have multiple organ failure, or a catastrophic antiphospholipid syndrome. This is caused by widespread microthrombi in multiple vascular beds, and can be devastating.

II. CASE REPORT

A 45-year-old lady called her GP complaining a 7-10 day history of swelling and pain in her right leg. There was no redness or warmth. Due to the pandemic a video consultation was arranged which confirmed that whole of right leg is swollen when compared to left. On further questioning denied any chest pain, mentioned about getting SOB lately. No Covid symptoms. No haemoptysis. Was referred to be hospital to exclude DVT and PE

She has had 2 c-sections without any VTE either during preg or during post partum period. Her mother had blood clot at age of 23 yr and no cause was found. This was around 50 yr ago and no further details was available. She used to work in a factory until around 2 yrs ago when she had an epileptic fit whilst at work. She is overweight and her BMI is 53. She is diabetic on insulin, she is on epileptic medication. Also PMH of Umbilical hernia and gall stones.

CTPA –bilateral extensive emboli extending to almost all the lobar and segmental branches. Evidence of right ventricular strain.

She received thrombolysis with alteplase successfully at the hospital.

USS Doppler right leg – the right common femoral vein was not patent or compressible with thrombus noted within.

Bloods on admission Hb105 MCV-73, PLT-315, Biochemical screen normal ferritin 35, b12-547. Cardiolipin antibodies Ab IgM-50. CardiolipinAbIG 7.5, HbA1C100

Due to the life threatening thrombotic event, a work-up for hypercoagulable state was initiated. Antinuclear antibody (ANA), rheumatoid factor (RF), factor V Leiden mutation, homocysteine level, protein C, protein S, C3, and C4 levels were all found to be within normal limits.

USS pelvis –subserosal fibroid. CT Abd pelvis- fibroid, cholelithiasis, umbilical and infraumbilical hernia

Echo- dilated right ventricle with impaired radial function, estimated PASP-36 MMHG+RA pressure, suggestive of Pulmonary HTN



III. DISCUSSION

Overweight and obesity are established risk factors for venous thromboembolism (VTE).

In one study PAPS was diagnosed in 6.8% (24/355) of normal weight (BMI < 24 kg/m²) VTE patients,

in 11.1% (50/452) of overweight (BMI 25-30 kg/m²) VTE patients,

and in 15.7% (30/191) of obese (BMI > 31 kg/m²) VTE patients.

The difference of PAPS occurrence between these groups was statistically significant (P = 0.001). PAPS patients demonstrated higher fibrinogen levels as compared to non-PAPS patients (median 416.0 mg/dl vs. 352.0 mg/dl, P = 0.001). Furthermore, fibrinogen levels increased significantly according to the body weight of patients (median normal weight patients 330.0 mg/dl vs. overweight patients 359.0 mg/dl vs. obese patients 415.0 mg/dl, P = 0.001).

IV. CONCLUSION

PAPS seems to be more frequent in overweight and obese patients. As PAPS patients showed significantly higher fibrinogen levels and as fibrinogen levels increased significantly according to the body weight of patients, an elevated inflammatory state in overweight and obese patients as a reason for the increased PAPS occurrence can be assumed.

Optimally, testing should be performed when a patient is clinically stable and not during an acute event. Interpretation of the results of laboratory tests performed near the time of an acute thromboembolic event can be difficult, as acute phase reactants such as factor VIII and fibrinogen may be markedly increased during acute events, altering coagulation test results. In addition, acute events may also trigger the appearance of anticardiolipin antibodies which are transient. Consequently, it is recommended that all positive results should be confirmed by repeat testing, at least twelve weeks apart, to distinguish patients with persistent antiphospholipid antibodies from those with transient antibodies. As in all subjects with thrombosis, attention should be paid to modifiable risk factors such as smoking, obesity and exogenous female hormone use. Although there is developing interest in, and some rationale for, use of alternatives to anticoagulant drugs to reduce thrombosis risk in APS, specifically statins (Ferrara et al, 2003, 2004) and hydroxychloroquine (Edwards et al, 1997; Espinola et al, 2002;

and et al, 2008), their use remains experimental at present

REFERENCES:

- [1]. Hughes GR. The anticardiolipin syndrome (editorial). *Clin Exp Rheumatol* 1985; 3: 285-6.
- [2]. Harris EN, Huges GR, Gharavi AE. Antiphospholipid antibodies: an elderly statesman dons new garments. *J Rheumatol* 1987; 13 (Suppl. 14): 208-13.
- [3]. World Health Organisation. Cardiovascular disease and steroid hormone contraception: report of a WHO scientific group. *WHO Tech Rep Ser* 1998; 877: 1-89.
- [4]. Farley TMM, Collins J, Schlesselman JJ. Hormonal contraception and risk of cardiovascular disease: an international perspective. *Contraception* 1998; 57: 211-30.
- [5]. Brown MD, Rowe BH, Reeves MJ, et al. The accuracy of the enzyme-linked immunosorbent assay D-dimer test in the diagnosis of pulmonary embolism: a meta-analysis. *Ann Emerg Med* 2002; 40: 133-44.
- [6]. Maloba M, Hogg K. Diagnostic utility of arterial blood gases for investigation of pulmonary embolus. *Emerg Med J* 2005; 22: 435-6.
- [7]. Wilson WA, Gharavi AD, Koike T, et al. International consensus statement on preliminary classification criteria for definite antiphospholipid syndrome: report of an international workshop. *Arthritis Rheum* 1999; 42: 1309-11.
- [8]. Neville C, Rauch J, Kassis J, et al. Thromboembolic risk in patients with high titre anticardiolipin and multiple antiphospholipid antibodies. *Thrombosis Hemostasis* 2003; 91: 108-15.
- [9]. Petri M. Epidemiology of the antiphospholipid antibody syndrome. *J Autoimmun* 2000; 15: 145-51. 10. McClain MT, Arbuckle MR, Heinlen LD, et al. The prevalence, onset and clinical significance of antiphospholipid antibodies prior to diagnosis of systemic lupus erythematosus. *Arthritis Rheumatism* 2004; 50: 1226-32.
- [10]. Chandrashekhara S, Kirthi R, Varghese J. Prevalence of anticardiolipin antibodies in various thrombotic conditions: a hospital-based study. *J Assoc Physicians India* 2003; 51: 359- 62