Case Report

A "Primary" Thrombotic Syndrome: Absence of Anti-Phospholipid Antibodies

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Summary

A patient presenting fetal loss, livedo reticularis, severe migraine crises, myocardial infarction and thrombotic vasculopathy of both external iliac arteries is described. The serologic study showed absence of antiphospholipid antibodies (aPL), suggesting that in some cases the presence of these antibodies may be a consequence of tissue damage.

Key words: Thrombosis, Antiphospholipid Antibodies, Anticardiolipin Antibodies.

INTRODUCTION

Anticardiolipin antibodies (aCL) and the lupus anticoagulant (LA) have been associated with a thrombotic diathesis in SLE, related autoimmune disorders, and other unrelated diseases (1).

Clinically, aPL have been associated with venous and arterial thrombosis, thrombocytopenia, haemolytic anaemia, livedo reticularis, recurrent fetal loss, chorea, thrombotic stroke and occlusion of coronary artery bypass grafts (2,3).

Due to the crescent importance attributed to these antibodies in the above cited manifestations, the present report describes a patient that fulfills the clinical criteria for a primary antiphospholipid syndrome without aPL.

CASE REPORT

A 37-year-old white woman was admitted to the Intensive Care Unit (ICU) with a three-hour history of severe chest pain that started during a period of exertion. She has never had angina before and there was no history of previous cardiac or pulmonary disease. She smoked about 10 cigarettes a day and rarely drank alcohol. Physical examination on admission was essentially normal except for the presence of livedo reticularis on both legs and abdomen. Blood pressure was 100/50 mm Hg and pulse rate 54 beats/min. An electrocardiogram demonstrated a first degree atrioventricular block, with frequent premature ventricular contractions and depressions of ST segments from V1 to V4. Diagnosis of acute infero-latero-dorsal myocardial infarction was confirmed also by elevation of cardiac isoenzymes.

Immediately after admission, recombinant tissue plasminogen activator (rTPA) was administered (100 mg/iv in 3 hours) and the patient had her haemodynamic parameters measured by thoracic electrical bioimpedance. Low cardiac output and an increased afterload were detected requiring the introduction of dobutamine and nitroprusside. After one day the patient was much better and vasoactive drugs could be discontinued. On the seventh day after admission, the patient was discharged from the ICU to the infirmary and six days later an echocardiogram was performed showing no thrombus and no significant motional abnormality.

Nine days after being discharged from the ICU the patient was readmitted because of a very strong pain in both legs that had started slightly the day before and became much worse, two hours before admission. The patient was very anxious, sudoretic and hyperventilating. Physical examination revealed pulseless bilateral legs that were pale and cold. Arteriography showed a normal aortic bifurcation. Both common and internal iliac arteries were also normal. The right and left external iliac arteries were not opacified over their entire length (Fig. 1).

Relevant laboratory results included: Hb 15.1 g/dl; WBC 13,400/mm³; platelets 503,000/mm³; prothrombin activity 100%; activated partial thromboplastin time 50 seconds (normal 35-50 seconds); fibrinogen 596 mg%; fibrin degradation products 23.5 mcg/ml (normal 2.5 - 5 mcg/ml); sedimentation rate 100 mm at one hour; C reactive protein positive at 1:320 dilution; total choles-
terol 145 mg/dl (HDL 35, LDL 37, VLDL 73); pH 7.5; bicarbonate 23 meq/l.

At this time, upon questioning, patient admitted having one spontaneous abortion at the fifth month of pregnancy, three years previously and several severe migraine crises. rTPA was again administered, followed by three daily shots of methylprednisolone. Forty-eight hours after therapy initiation, femoral and popliteal pulses were noted bilaterally. One week later the patient recovered her normal arterial status and was asymptomatic under prescription of oral corticosteroid and platelet aggregation antagonists.

Her frozen serum collected at the time of her second admission was used for the performance of the following laboratory tests: aCL, LA (APTT Mix and ELISA); anti-neutrophil cytoplasmic autoantibodies (ANCA), and antinuclear antibodies (ANA). None of the above-cited autoantibodies showed a positive result.

**DISCUSSION**

The presence of aPL may be a serologic marker for the thrombotic vasculopathy occurring in SLE and lupus-like diseases.

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**REFERENCES**