Cardiovascular risk in rare autoimmune diseases, lessons from a primary antiphospholipid syndrome patients

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Background: Antiphospholipid syndrome is a thrombo-inflammatory disease driven by circulating autoantibodies (aPL) that recognize cell surface phospholipids and phospholipid-binding proteins. In the absence of an underlying connective tissue disorder, the persistent presence of these antibodies associated with recurrent fetal losses and arterial or venous thrombosis is defined as a primary antiphospholipid syndrome (PAPS) with a prevalence of 1/2000 people, predominantly female.

Purpose: Estimation of cardiovascular risk presence based on markers of subclinical cardiovascular impairment.

Methods: 100 PAPS patients, with an average age of 47.70±13.14y, 93.7% females, were included in the study. Anticardiolipin antibodies (aCL IgG/IgM), anti-ß2 glycoprotein-I (anti-ß2GPI IgG/IgM), and lupus anticoagulant (LAC) were determined. Abnormal cut-off values used for left ventricular diastolic dysfunction (LVDD) were septal $E' < 7$ cm/sec, lateral $E' < 10$ cm/sec, average $E/E’$ ratio $>14$, LA volume index (LAVI) $>34$ mL/m², and peak tricuspid regurgitation velocity $>2.8$ m/sec. LVDD was present if more than half parameters were with abnormal values. Endothelial dysfunction was assessed through ultrasound measurement of brachial artery flow-mediated dilation (FMD) and atherosclerosis progression by ultrasound assessment of intima-media thickness (cIMT) on carotid arteries (common carotid artery, CCA, internal carotid artery, ICA). The results were compared to 90 healthy, age and sex-matched controls.

Results: LVDD was significantly more prevalent in PAPS patients compared to healthy controls (24.8% vs. 2.2%, p=0.001). FMD was significantly lower in PAPS patients (10.76±5.15% vs. 14.20±6.76%, p=0.0001) and cIMT values were significantly higher (left ICA 0.80±0.29mm vs 0.56±0.20mm, p=0.001). All markers of subclinical atherosclerosis were significantly related to age and standard atherosclerotic risk factors present. Regarding aPL profile, anti-ß2GPI IgG positive PAPS patients had higher levels of cIMT ICA left (p=0.049) and LAC positive higher prevalence of LVDD (p=0.047) CONCLUSIONS: PAPS patients impose significantly higher prevalence of subclinical cardiovascular involvement compared to healthy population. Timely recognition, along with vigorous control of standard atherosclerotic risk factors and a proper therapeutic (antithrombotic and/or anticoagulant) regime, is of great importance.