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The treatment of anti-phospholipid syndrome: A comprehensive clinical approach

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ABSTRACT

Anti-phospholipid syndrome (APS) is an acquired pro-thrombotic autoimmune disease that predisposes to thrombotic events and/or obstetric complications, in the persistent presence of anti-phospholipid antibodies (aPL). Life long moderate-intensity anticoagulation is the option of choice for aPL-positive patients with a previous thrombosis; critical issues concern the management of those with a history of arterial event due to the high rate of recurrence. Alternatives comprise anti-platelet agents and high-intensity anticoagulation. Low dose aspirin (LDASA) and low molecular weight heparin provide the mainstay of the treatment of obstetric APS, allowing a birth rate in 70% of cases. The management of refractory APS, thrombotic as well as obstetric, is highly debated, but an increasing burden of evidence points towards the beneficial effects of multiple treatments. Similarly, a management envisaging multiple drugs (anticoagulation, steroids, plasma exchange and/or intravenous immunoglobulins) is the most effective approach in catastrophic APS. Asymptomatic aPL carriers are at higher risk of thrombotic and obstetric complications compared to the general population, thus potentially benefitting of a pharmacological intervention. LDASA and hydroxychloroquine can be considered as options, in particular in case of high risk aPL profile, concomitant cardiovascular risk factors or associated autoimmune disease. APS is apparently a simple condition, but its multifaceted nature requires a complex and tailored treatment.

Contents

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1. Introduction

Anti-phospholipid syndrome (APS) is an acquired pro-thrombotic autoimmune disease that predisposes to thrombotic events and obstetric complications, potentially manifesting at any gestational age, in the persistent presence of anti-phospholipid antibodies (aPL). Among pro-thrombotic conditions, APS is unique as thrombosis might happen at any site throughout the whole vascular three: veins, arteries and microvascular system. Multiple thromboses might present concomitantly in different vascular districts, configuring the catastrophic variant of the syndrome (CAPS). These clinical manifestations are enlisted in the updated classification criteria for APS

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