XXV Congresso de Iniciação Científica da Unicamp

October 18 to 20 Campinas | Brazil

2017

Prognostic impact of antiophospholipid antibodies on the clinical course of primary immune thrombocytopenia or autoimmune hemolytic anemia

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Abstract

This project has the aim to evaluate the influence of antiphospholipid antibody (aPL) in the clinical course of Immune Thrombocytopenic Purpura (ITP) and Autoimmune Haemolytic Anemia (AHA), since it is controversial if ITP and AHA should be defined as Antiphospholipid Syndrome (APS).

Key words:

Antiphospholipid antibodies, primary immune thrombocytopenia, autoimmune hemolytic anemia.

Introduction

This project has the aim to evaluate the influence of antiphospholipid antibody (aPL) in the clinical course of Immune Thrombocytopenic Purpura (ITP) and Autoimmune Haemolytic Anemia (AHA), since it is controversial if ITP and AHA should be defined as Antiphospholipid Syndrome (APS).

In a prospective cohort study, patients with primary ITP and AHA were enrolled between January 2013 and January 2016 and followed up until December 2016.

Lupus anticoagulant (LAC), IgG/IgM anticardiolipin and anti-beta2-glycoprotein1 (aB2GP-1) assays were performed at diagnosis. The primary endpoint was the response to first-line treatment and the secondary endpoints were multiple treatment lines, splenectomy, thrombosis and bleeding events.

Results and Discussion

Eighty-four patients (72 ITP, 11 AHA, 1 Evans syndrome) were included and followed for 17 months (3.5 - 38 months). Twenty-two patients (26.2%) were positive for aPL (10 LAC, 7aB2GP-1 and 5 double-positive).

aPL-positive patients were predominantly female (77.3% vs. 53.2%, $P=0.04$) and 90.9% met the criteria for initial treatment (versus 85.5% of aPL-negative patients, $P=0.5$). The response to first line-treatment was similar between groups (75% in aPL-positive vs. 70.5% in aPL-negative; $P=0.5$). Cox regression analysis revealed that aPL positivity was not a risk factor for first-line treatment failure ($HR=1.38$ 95%CI=0.69-2.75, $P=0.3$), multiple treatment lines ($HR=1.30$ 95%CI=0.66-2.59, $P=0.4$) and splenectomy ($HR=0.88$ 95%CI=0.26-3.08, $P=0.8$). Four patients had thrombosis and aPL positivity was not a risk factor for thrombosis ($HR=0.65$ 95%CI=0.09-4.69, $P=0.6$). The risk for bleeding events was also similar between positive and negative aPL patients ($HR=1.55$ 95%CI=0.82-2.93, $P=0.2$).

Figure : Theses figures illustrate the cumulative risk of thrombosis and bleeding during the follow-up. The risks were similar between patients with positive aPL and negative aPL

Conclusions

The results demonstrated that aPLs do not influence the clinical course of primary ITP or AHA. The study suggests that aPL-positive ITP or AHA are not distinct entities, not supporting the inclusion of these aPL-related manifestations as clinical criteria for APS diagnosis.

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DOI: 10.19146/pibic-2017-78270

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